

VOICE OF THE PATIENT REPORT



HLAA's Externally-Led Patient-Focused Drug Development (PFDD) Meeting for People and Families Living with Sensorineural Hearing Loss

Virtual Meeting Held May 25, 2021
10 a.m. - 3 p.m. Eastern Time
Report Date: September 30, 2021



HLAA's Externally-Led Patient-Focused Drug Development (PFDD) Meeting for People and Families Living with Sensorineural Hearing Loss

Meeting hosted by: Hearing Loss Association of America

Submitted to: Office of Tissues and Advanced Therapies, Center for Biologics Evaluation and Research, U.S. Food and Drug Administration (FDA).

The "Voice of the Patient" report and the meeting recording are available at hearingloss.org/programs-events/patient-focused-drug-development-meeting/.

This meeting was made possible, in part, with support provided by the following companies:



Partners

Thank you to these organizations and companies who support hearing health care research and solutions.

- Academy of Doctors of Audiology
- The Alexander Graham Bell Association for the Deaf and Hard of Hearing (AG Bell)
- Alport Syndrome Foundation
- American Academy of Audiology
- American Academy of Otolaryngology-Head and Neck Surgery (AAO-HNS)
- AARP
- American Speech-Language-Hearing Association (ASHA)
- American Tinnitus Association
- Association of Adult Musicians with Hearing Loss
- Association of Late-Deafened Adults (ALDA)
- Bionics Institute
- British Cochlear Implant Group
- Canadian Hard of Hearing Association (CHHA)
- Canadian HARD of HEARING Association (CHHA-Hamilton)
- CaptionCall
- Cochlear
- Cochlear Implant International Community of Action (CIICA)
- Deafness Forum of Australia
- Deafness Foundation Australia
- Ear Community
- European Association of Cochlear Implant Users (EURO-CIU)
- European Federation of Hard of Hearing People (EFHOH)
- The Global Alliance of Speech-to-Text Captioning
- Hands & Voices
- Hearing Health Foundation (HHF)
- Hearing Industries Association
- The Hearing Journal
- Hearing Matters Australia
- Hearing Rehabilitation Foundation
- Hearing Tracker
- Ida Institute
- International Federation of Hard of Hearing People (IFHOH)
- International Hearing Society (IHS)

Voice of the Patient Report: *Living with Sensorineural Hearing Loss*

- MED-EL Corporation
- National Guard Association of the United States (NGAUS)
- Navy SEAL Foundation
- NIHR University College London Hospitals Biomedical Research Centre
- New York University Grossman School of Medicine
- Oticon, Inc.
- OTOjOY
- Phonak
- Pindrop Foundation
- ReSound US
- Rexton
- RNID-UK
- Sharon L. King Foundation
- Signia
- Starkey
- Tennessee Captioning
- Tinnitus Hub
- U.S. Department of Veterans Affairs
- U.S. Special Operations Command (SOCOM)
- University of Iowa Department of Otolaryngology Cochlear Implant Clinical Research Centre
- Usher Syndrome Coalition
- Vanderbilt Bill Wilkerson Centre for Otolaryngology and Communication Sciences
- Widex

Collaborator

- Department of Defense Hearing Center of Excellence

Voice of the Patient Report: *Living with Sensorineural Hearing Loss*

The Hearing Loss Association of America (HLAA) is the leading organization representing consumers with hearing loss. The mission of HLAA is to open the world of communication to people with hearing loss by providing information, education, support and advocacy. This “Voice of the Patient” report was prepared by HLAA as a summary of the input shared by people and families living with Sensorineural Hearing Loss (SNHL) during an Externally-Led Patient-Focused Drug Development Meeting (EL-PFDD). This hearing-accessible meeting was hosted virtually on May 25, 2021.

Authors and Collaborators

This report was prepared and submitted on behalf of HLAA by Barbara Kelley, executive director, HLAA; Carrie Johnson, director of communications, HLAA; and by Chrystal Palaty, medical writer.

Consulting partners include James Valentine, Esq., and Larry Bauer, R.N., M.A., from Hyman, Phelps & McNamara, P.C.

Disclosures: HLAA is a tax-exempt, charitable organization and is eligible to receive tax-deductible contributions under IRS Code 501(c)(3). HLAA receives funding from (pharmaceutical/medical device/technology) companies in the form of unrestricted grants and sponsorship of programs and events. HLAA contracted with Chrystal Palaty from Metaphase Health Research Consulting Inc. for assistance in writing the report. James Valentine and Larry Bauer are employed by Hyman, Phelps & McNamara, P.C., a law firm that represents companies who are developing therapeutics and technologies to advance health, as well as patient advocacy organizations.

Technical services: Provided by John Dudley, Eric Quigley and Kyle Rhoderick of Dudley Digital Works.

Funding received for the Sensorineural Hearing Loss Externally-Led PFDD meeting included financial support from: Acousia Therapeutics, Akouos, Decibel Therapeutics, Frequency Therapeutics, Otonomy, Pipeline Therapeutics and Sensorion. In return for monetary support, these companies were acknowledged at the beginning of the meeting and their respective logos were displayed during the meeting break. These supporters did not have input in design, planning, coordination or execution of the meeting.

Report Version Date: September 30, 2021

Revision statement: This document was not revised and/or modified in any way after September 30, 2021.

Statement of use: HLAA have the necessary permissions to submit “Voice of the Patient Report: Living with Sensorineural Hearing Loss” to the FDA. Linking to this resource from the FDA website does not violate the proprietary rights of others. Permission to link from the FDA website is granted by HLAA (hearingloss.org).

Voice of the Patient Report: *Living with Sensorineural Hearing Loss*

Point of Contact: For questions related to this report please contact Barbara Kelley, executive director, HLAA at bkelly@hearingloss.org.

Acknowledgements

The Hearing Loss Association of America sincerely thanks the people with hearing loss who shared their personal stories and insights at this meeting, to help everyone to better understand hearing loss and its impacts. We thank the panelists who spent many hours preparing their statements and the discussion starters for sharing their experiences. We also want to thank everyone who called or wrote in for providing their insights and comments. Without you, this meeting would not have achieved its goals.

Our most sincere appreciation goes to the U.S. Food and Drug Administration for permitting HLAA to hold this meeting and to the many FDA staff members who made the time to attend. Thank you to William Lewellen FAC-P/PM, from the FDA, who guided HLAA through this process. Thanks also to the many representatives from the military, government agencies, state agencies, academia, nonprofit organizations and industry who attended and listened.

We thank Gavin Imperato, M.D., medical officer, from the Office of Tissues and Advanced Therapies of the FDA for his informative opening remarks. We thank Frank Lin, M.D., Ph.D., director, Cochlear Center for Hearing and Public Health and professor of Otolaryngology—Head and Neck Surgery at Johns Hopkins Bloomberg School of Public Health, for providing a clear clinical overview of hearing loss. We are grateful to you both for supporting us with your time and expertise.

Thank you to everyone whose work ensured that our voices were heard. Thank you to Dudley Digital Works, our media team, captioners, sign language interpreter and to James Valentine and Larry Bauer. We wish to thank the HLAA staff who worked hard to plan this meeting.

HLAA wishes to thank our generous supporters who financially sponsored this meeting including Acousia Therapeutics, Akouos, Decibel Therapeutics, Frequency Therapeutics, Otonomy, Pipeline Therapeutics and Sensorion. Our sincere appreciation goes to you.

Finally, we wish to acknowledge our 56 partner organizations and our collaborator for this meeting including: advocacy and professional organizations, companies, federal agencies and universities from across the world who support hearing health care research and solutions. Their commitment to their work is a testament to the enormity of the importance of hearing loss as a global public health issue.

Voice of the Patient Report: *Living with Sensorineural Hearing Loss*

Contents

Executive Summary	7
Introduction and Meeting Overview	8
Overview of Sensorineural Hearing Loss	8
Meeting Summary.....	9
Report Overview and Key Themes.....	11
Topic 1: Symptoms and Daily Impacts of Hearing Loss	14
Physical health concerns experienced by people with hearing loss include trouble hearing with background noise, fatigue and sounds are muffled	14
People with hearing loss also experience many psychosocial health concerns such as social isolation, depression and anxiety, verbal communication issues	18
People with hearing loss described how hearing loss significantly impacts all aspects of their lives....	21
People with hearing loss have many worries for the future including increased risk for dementia.....	29
Topic 2: Current and Future Treatments	31
Different situations need different solutions: people with hearing loss use a combination of technology solutions, medical treatments or medications to address their hearing loss.....	31
Most poll respondents use multiple additional approaches to address hearing loss and manage symptoms.....	34
Most meeting participants reported that their current regimen controls their condition “to a great extent” or “somewhat” but identified many drawbacks.....	39
People with hearing loss prioritized hearing restoration and improved hearing as most important for a possible new drug, device or treatment.....	42
Incorporating Patient Input into a Benefit-Risk Assessment Framework	48
Conclusion	51
Appendix 1: Meeting Demographics	52
Appendix 2: Meeting Agenda	55
Appendix 3: Meeting Discussion Questions	56
Appendix 4: Panel Participants and Callers	57
Appendix 5: Topic 1 Polling Results. Symptoms and Daily Impacts of Sensorineural Hearing Loss	58
Appendix 6: Topic 2 Polling Results. Current and Future Approaches to Treatment for Sensorineural Hearing Loss	61

Voice of the Patient Report: *Living with Sensorineural Hearing Loss*

Executive Summary

The Hearing Loss Association of America (HLAA) held an Externally-Led Patient-Focused Drug Development Meeting (EL-PFDD) for people and families living with Sensorineural Hearing Loss (SNHL) on Tuesday, May 25, 2021. The meeting brought together people with SNHL and their families, representatives of the FDA, government agencies and the military, as well as representatives from state agencies. Meeting attendees included representatives from industry, academia, advocacy groups and nonprofit organizations; in total, seven supporters, 56 partners and one collaborator joined HLAA to support this meeting.

This EL-PFDD meeting was modeled after the work of the FDA's Patient-Focused Drug Development (PFDD) initiative. PFDD is a systematic way of gathering patient perspectives on their condition and on available treatments. The information gathered at the meeting is presented in this "Voice of the Patient" report. The information in this report will be used to guide drug and technology development and inform the FDA's benefit-risk evaluations when reviewing technologies and therapeutics to address hearing loss.

Sensorineural hearing loss (SNHL) results from damage to the hair cells or nerve fibers of the inner ear that convert sound into electrical impulses. Sensorineural hearing loss is not just a reduction of sound level and degradation of speech perception but includes a very complex and interfering combination of physical and psychosocial health concerns. There is no such thing as a "small" hearing loss and each hearing loss is unique with varying consequences and effects.

People with hearing loss described the impacts of hearing loss. Hearing loss is an invisible disability, is heavily stigmatized and leads to social isolation and a diminished quality of life. The strong link between hearing loss and dementia is a worry for people with hearing loss. Many expressed worries of being further isolated due to losing social connections and relationships, losing the ability to communicate and losing additional hearing. Those with hereditary hearing loss expressed worries for their children and grandchildren.

There is a continued unmet need for solutions to support people with hearing loss. People with SNHL use combinations of technologies, tools, medical treatments, accommodations and strategies to address their hearing loss. While most of these tools help to amplify sound and/or reduce symptoms of hearing loss, they are not able to replace hearing. Many commented on the high cost of hearing solutions, especially as many accommodations are required. While disease modifying or curative therapies for hearing loss are not yet available, people with hearing loss are hopeful that the future will provide better solutions that might restore or improve their hearing.

The hope is that the information obtained at this event will ultimately catalyze significant improvements for the health and quality of life for people living with SNHL.

Voice of the Patient Report: *Living with Sensorineural Hearing Loss*

Introduction and Meeting Overview

The Hearing Loss Association of America (HLAA) received permission from the U.S. Food & Drug Administration (FDA) to hold an Externally-Led Patient-Focused Drug Development Meeting (EL-PFDD) for people and families living with Sensorineural Hearing Loss (SNHL) on Tuesday, May 25, 2021. While this meeting was held virtually for participants' health and safety due to the COVID-19 pandemic, the virtual platform allowed increased participation across the United States and the world. The meeting was hearing accessible and included real-time captioning and American Sign Language (ASL) interpretation.

This EL-PFDD meeting was modeled after the work of the FDA's Patient-Focused Drug Development (PFDD) initiative. PFDD is a systematic way of gathering patient perspectives on their condition and on available treatments. The information gathered at the meeting is presented in this "Voice of the Patient" report. The information in this report will be used to guide drug and technology development and inform the FDA's benefit-risk evaluations when reviewing technologies and therapeutics to address hearing loss. The hope is that this information will ultimately catalyze significant improvements for the health and quality of life for people living with SNHL. In this report, we refer to "SNHL" and "hearing loss" interchangeably.

Overview of Sensorineural Hearing Loss¹

Sensorineural hearing loss (SNHL) results from damage to the hair cells or nerve fibers of the inner ear that convert sound into electrical impulses. These hair cells are critical for hearing sound as well as the tuning and filtering of sound. SNHL is a permanent and degenerative condition that encompasses noise-induced and age-related hearing loss, as well as hearing loss caused by ototoxic medications, some viruses and idiopathic sudden hearing loss. SNHL can be heritable, as a result of DFNA5 or GJB2 gene defects, and may present as part of other disorders such as Alport syndrome² or Ménière's disease. No matter the cause, SNHL results in decreased sound sensitivity and distortion in sound encoding resulting in a garbled sound signal. Often words and speech cannot be discriminated, leading to the common concern of "*I can hear but I can't understand.*"

SNHL loss is the cause of more than 90% of all hearing loss, impacting tens of millions of people in the United States. Currently, 48 million people in the United States experience hearing loss

¹ Summary extracted from the original Letter of Intent submitted to the FDA and from the Clinical Overview of Hearing Loss provided by Frank Lin, M.D., Ph.D. Director, Cochlear Center for Hearing and Public Health, Professor of Otolaryngology—Head and Neck Surgery, Johns Hopkins Bloomberg School of Public Health.

² Alport syndrome, a rare genetic kidney disease, also causes bilateral SNHL. The Alport Syndrome Foundation held an EL-PFDD meeting on August 18, 2018, and many of their patient comments reflect the comments from this meeting. Their report is available on their website. <https://www.alportsyndrome.org/about-alport-syndrome/voice-of-the-patient-report/>.

Voice of the Patient Report: *Living with Sensorineural Hearing Loss*

and this is expected to almost double, to 73.5 million by 2060. Age is the biggest predictor of hearing loss, with hearing loss doubling with every decade of life. SNHL significantly impacts individuals who are exposed to hazardous noise in the workplace, including those in the military, in manufacturing and in industry.

Hearing loss was identified as a “primary health concern” by The National Academies of Sciences, Engineering, and Medicine’s study on [Hearing Health Care for Adults: Priorities for Improving Access and Affordability](#), for which HLAA was the only consumer-group sponsor. Beyond the condition of SNHL itself, an array of health issues may arise from even mild forms. Hearing loss in mid and late life as being single-handedly the largest risk factor for dementia of all known risk factors, as determined by a large systematic meta-analytic review by the [Lancet Commission on Dementia](#). In addition to dementia and brain aging, hearing loss in adults is increasingly identified as an independent risk factor for health care expenditures/utilization, impaired physical functioning, the risk of progressing to a disability, a fall or needing nursing care. People with hearing loss have a greater risk of needing an emergency room visit, of having a longer hospital stay or a readmission to the hospital over time, as well as a greater rate of hospitalization over time. Hypothesized mechanisms for how hearing loss contributes to dementia include cognitive load, brain structural changes (atrophy), social isolation or loss of environmental sound cues.

At this time, there are no medical or surgical solutions to address SNHL or its underlying causes and damage is irreversible. Current management options include clinic-based auditory rehabilitation (auditory needs assessment, communication strategies) and sensory management with hearing aids, hearing assistive technologies and cochlear implants for those with more severe hearing loss. Novel approaches under development for hearing loss management include alternative service delivery models such as telehealth, direct consumer access to safe and effective over-the-counter hearing aids, and innovations in amplification strategies, specifically the development of better algorithms for manipulating auditory signal through a hearing aid or cochlear implant. While not yet available, gene therapies and regenerative medicine approaches are currently being developed and evaluated and represent a promising paradigm shift in SNHL treatment. These approaches could potentially be neurorestorative rather than rehabilitative.

Meeting Summary

The May 25, 2021 “Living with Sensorineural Hearing Loss” EL-PFDD meeting was the 47th EL-PFDD and due to the ongoing COVID-19 pandemic, was the 14th fully virtual EL-PFDD meeting. While this meeting was held virtually for participants’ health and safety due to the COVID-19 pandemic, the virtual platform increased accessibility by enabling participation from across the United States and the world. The meeting was hearing accessible and included real-time captioning and ASL interpretation.

Voice of the Patient Report: *Living with Sensorineural Hearing Loss*

The demographics of the meeting attendees living with hearing loss was determined using online polling and are summarized below as well as in **Appendix 1**. Most poll respondents, 86%, were individuals living with hearing loss while the remaining 14% were relatives or care partners of someone with hearing loss. Most poll respondents, 92%, were located in the continental U.S., with a few representatives from Canada, Europe, Middle East and other countries. A total of 68% attendees were female, 31% were male and 1% were non-binary. Although attendees spanned the entire adult age range, the greatest representation of attendees were 71 years of age or older (36%), where the incidence of hearing loss is the highest, and more than 50% were older than 61 years of age. Attendees reported first experiencing symptoms of hearing loss across the age spectrum, however 44% reported that they experienced their first symptoms of hearing loss early in life, from birth to age 18, with a decrease for each age interval.

Most attendees, 97%, reported hearing loss in both ears, with the remaining 3% who had hearing loss in the left ear only. Attendees reported a range of different forms of hearing loss, with 36% experiencing genetically-based or hereditary hearing loss, 21% with hearing loss of unknown origin, 10% from post loud noise exposure, 8% after a disease or virus, 6% unsure of the cause, 6% sudden onset, 5% aging, 3% tumor, 3% Ménière's disease, 1% autoimmune and 1% post head or ear trauma. Although none of the polling respondents reported experiencing hearing loss due to medication toxic to the ear, one of the Zoom panelists did report having hearing loss as a result of chemotherapy.

In addition to people living with hearing loss and their family members, caregivers and friends, the meeting was attended by representatives of the FDA, government agencies and the military (45), as well as representatives from state agencies. Meeting attendees included representatives from industry (79), academia (3), advocacy groups and nonprofit organizations (20).

The Living with Sensorineural Hearing Loss (EL-PFDD) meeting was structured around two main topics. The morning session was structured around Topic 1, *Symptoms and Daily Impacts of Living with Sensorineural Hearing Loss*, while the afternoon session addressed Topic 2, *Current and Future Treatment for Sensorineural Hearing Loss*. The meeting agenda is in **Appendix 2**, and the questions provided for meeting discussion are in **Appendix 3**.

The meeting was opened by Barbara Kelley, executive director of HLAA, who welcomed and thanked all attendees for their participation. Gavin H. Imperato, M.D., medical officer, Office of Tissues and Advanced Therapies, Center for Biologics Evaluation and Research, U.S. Food and Drug Administration, welcomed meeting participants and described the FDA's role throughout the entire drug development process and emphasized how important patient voices are in this process. He reminded attendees that the FDA consists of human beings who are themselves patients, family members and friends and are personally affected by so many of the diseases and conditions that they regulate. He closed by thanking all the attendees for their courage and

Voice of the Patient Report: *Living with Sensorineural Hearing Loss*

candor and stated that their participation is absolutely critical, especially when thinking about the safety and efficacy framework of new therapeutics. Frank Lin, M.D., Ph.D., director, Cochlear Center for Hearing and Public Health, Professor of Otolaryngology—Head and Neck Surgery, Johns Hopkins Bloomberg School of Public Health, provided a scientific and medical overview of hearing loss.

The morning session continued with five panelists who each spoke about their experience with hearing loss. These panelists were selected to represent a range of hearing loss experience, and described their stories of discovering their hearing loss, the health concerns they experience, the impacts that hearing loss has had on their lives and their hearing loss-related worries for the future. Meeting attendees had the opportunity to call in, to submit online written comments and to participate in online polling. James Valentine moderated a discussion among several people who served on a Zoom panel and people who called in by phone. Additional relevant comments submitted through online comment submission form were read by Barbara Kelley. The first names of panelists and callers are listed in **Appendix 4**.

The afternoon session opened with a Zoom panel of five people living with hearing loss who described different technologies and other approaches they use to address their hearing loss challenges. Panelists described successes as well as the many challenges of unmet treatment needs. Again, meeting attendees were provided with an opportunity to have their voices heard through online polling, by calling in and by submitting written comments about the issues discussed, and their voices were added to the moderated discussion by Barbara Kelley and James Valentine. The meeting concluded with a summary of the key meeting points by Larry Bauer and was closed by Barbara Kelley who thanked all the participants and attendees.

The results of online polling results for Topics 1 and 2 are included in **Appendices 5** and **6**, respectively. To include as many voices as possible, an online comment submission portal was open for 30 days before and after the meeting. Selected comments are included in the body of this report, and all submitted comments are included in a separate, online PDF. The PDF of submitted comments along with the final “Voice of the Patient” report, the meeting transcript and a recording of the meeting can be found at: hearingloss.org/programs-events/patient-focused-drug-development-meeting/. According to HLAA YouTube statistics, the meeting has been streamed 379 times (as of September 20, 2021).

Report Overview and Key Themes

This report provides a high-level summary of the perspectives generously shared by people and their families living with sensorineural hearing loss who participated in the May 25, 2021, EL-PFDD meeting, and includes selected comments submitted through the online portal. HLAA has

Voice of the Patient Report: *Living with Sensorineural Hearing Loss*

provided this report to the FDA, and it is publicly available for the many stakeholders in the hearing loss community, including governmental agencies, regulatory agencies, medical product developers, researchers and health care professionals.

The input received from the May 25, 2021, EL-PFDD meeting reflects a range of experiences with, and perspectives on hearing loss and hearing loss management approaches. We acknowledge that participants at this meeting does not fully represent the diverse population and the many varied perspectives of people with hearing loss. The intent of this meeting was to focus on the community that wanted to specifically address how treatments could improve their quality of life. There may be health concerns, impacts, treatments or other aspects of hearing loss that are not included in this document. The terms used in this report to describe specific symptoms and treatment experiences reflect the words and language used by meeting participants during the meeting and in submitted comments. The quotes that appear in this report were made by people with hearing loss and were selected to represent both the common experiences of many people with hearing loss, as well as some of the less common symptoms and experiences.

This report intends to identify themes that emerged directly from people and families living with hearing loss to create a better understanding of (a) the health concerns and burdens of hearing loss in daily life, both common and more rare experiences that exist, and (b) the challenges and trade-offs in selecting appropriate technologies or treatment, which highlights a massive unmet medical need for effective treatments for people living with hearing loss.

Key themes that emerged from the May 25, 2021, HLAA meeting on Sensorineural Hearing Loss:

- 1. Sensorineural hearing loss is not just a reduction of sound level and degradation of speech perception but includes very complex and interfering combination of physical and psychosocial health concerns.** The combinations and impacts of these health concerns are different for each individual.
- 2. There is no such thing as a “small” hearing loss.** Each hearing loss is unique with varying consequences and effects. Those with unilateral SNHL experience the same physical and psychosocial health concerns and the same impacts on activities of daily living and quality of life as those with bilateral SNHL.
- 3. Hearing loss is an invisible disability.** People with hearing loss are forced to negotiate a world where hearing loss accommodation is not recognized as essential. Insurance companies don’t consider hearing aids or other assistive technologies as medical necessities. Environments that are inaccessible include hospitals, airports, schools and workplaces.
- 4. Hearing loss is heavily stigmatized.** Many people try to hide their disability. Some described bullying, intentional exclusion, and misperceptions about their intelligence. Many were in denial or ashamed about their hearing loss, which made them reluctant to seek treatment.
- 5. Hearing loss leads to social exclusion and a diminished quality of life.** Hearing loss profoundly impacts all aspects of daily life including social and family relationships, employment, education, participation in socialization, recreational and sports activities. Hearing loss extracts a heavy emotional toll, and people described their feelings of loss, grief and frustration.
- 6. The strong link between hearing loss and dementia is a worry for people with hearing loss.** Many expressed a fear of being further isolated due to losing social connections, losing the ability to communicate, losing additional hearing and losing their relationships. Those with hereditary hearing loss expressed fears for their children and grandchildren.
- 7. People with hearing loss use “every tool in the box”: combinations of technologies, tools, medical treatments, accommodations and strategies.** While most of these tools help to amplify sound and/or reduce symptoms of tinnitus (for example), they do not replace hearing and are often costly.
- 8. Disease modifying or curative therapies for hearing loss are not yet available, yet people with hearing loss want better treatments and technologies for hearing loss.** Many attendees described how far technology has already advanced in their lifetimes, with improved (upgraded) hearing aid and cochlear implant technologies, speech-to-text apps. They are hopeful of new individualized therapies including the promise of regenerative medicine.

Topic 1: Symptoms and Daily Impacts of Hearing Loss

During the Sensorineural Hearing Loss EL-PFDD meeting, meeting attendees with hearing loss described their experience with hearing loss diagnosis and how they hear sound. A key point emphasized throughout the meeting was that hearing loss is not just a reduction in sound perception, but a loss of the ability to discriminate speech and involves a very complex and interfering combination of health concerns.

For Jake, a young adult with sudden hearing loss due to loud noise exposure at the age of 18, *“The symptoms I have experienced have stretched far beyond just the hearing damage and hyperacusis and tinnitus...I started developing vertigo, which turned into vestibular migraines, which have basically taken control of my life. My ear sensitivity causes migraines, which in turn makes my ear sensitivity and tinnitus even worse. All of this has caused major strife in my life and has made things much more complicated.”*

Kim., who experienced gradual one-sided hearing loss due to Ménière’s disease at the age of 40, said *“I certainly identified with pretty much all of the symptoms that were on the polling. I can say at one time or another, I have experienced some more than others because of the nature of Ménière’s.”*

Meeting attendees used online polling to identify all hearing loss-related health concerns that they had experienced as well as their top three most troublesome health concerns. Hearing loss-related health concerns fell into two categories: physical and psychosocial. They are listed below in the order of priority according to the online poll results and are presented in **Appendix 5, Q1** and **Q2**. Additional symptoms identified during the meeting and from online comments are also described.

Of note, people with one sided hearing loss experience all the same physical and psychosocial health concerns as those with bilateral hearing loss. Both Kim and Ali, who have single sided, viral-induced hearing loss, described experiencing the same health concerns and the same impacts as the rest of meeting attendees.

Physical health concerns experienced by people with hearing loss include trouble hearing with background noise, fatigue and sounds are muffled

Trouble hearing with background noise

Trouble hearing with background noise was the top health concern experienced by 92% of poll respondents and was selected as the most troublesome hearing loss-related health concern by 82% of respondents. Background noise was described by Tim, who experienced moderate bilateral hearing loss from birth, as an *“Overwhelming hearing environment.”*

Voice of the Patient Report: *Living with Sensorineural Hearing Loss*

Ron, who has gradual hearing loss due to Ménière's disease and/or acoustical trauma, shared his experience of hearing sound as noise. *"I can hear, I hear noise all the time. I just don't understand the words. So that's very, very frustrating."*

Latisha, born with hereditary SNHL described how much of a problem background noise is for her. *"Noise, it just blocks out any type of speech whatsoever. It's frustrating. It causes fatigue. It makes me tired. ... I just hear sound and noise and background noise makes it even harder for me to hear speech."*

Russ, who experienced progressive hearing loss for the last 25 years said, *"I'm defining noise pretty broadly to talk both about unwanted environmental noise, as well as speech that you really don't want to hear, because it interferes with what you do want to hear."*

Fatigue

While 62% of poll respondents experience hearing-loss related fatigue, 33% picked this as one of their top concerns. Fatigue was described many times during the meeting. For Linda, who experienced hearing loss during adulthood, *"The thing that I find to be a consistent problem, regardless of the technology that I use, is listening fatigue and just always being really, really tired."* She continued, *"That fatigue affects my work, it affects my relationships. It affects basically everything I do in my life."* Linda also described how fatigue increased as her ability to hear declined and how she, *"Had a limit on the number of hours I could listen in a day."*

Tim, who has moderate bilateral hearing loss from birth, described how, *"It takes a lot of mental energy for me to hear. Getting older, I find fatigue settles in more intensely. I can't enjoy my family and friends quite as much as I used to because I need more time to reset. It's sad to me, but Zoom gatherings, talking on the phone and social media just wears me out faster."*

Zina, diagnosed with hearing loss at the age of 3 1/2, described how *"COVID-19 significantly increased my listening fatigue. With face masks I cannot supplement my hearing with lipreading. To compensate the listening fatigue that I, and most people with hearing loss face, I take naps after class and go for walks."*

Sounds are muffled

A total of 66% of poll respondents experience this health concern, and 26% selected this as one of their top three most troublesome health concerns. For Cyleste, *"My main symptom is the inability to understand conversational speech in a call or Zoom meeting, in a group situation or with anyone wearing a mask."*

Joe described how his hearing loss has left him with, *"[The] inability to hear much language, music and nature sounds."*

Voice of the Patient Report: *Living with Sensorineural Hearing Loss*

This issue became much worse with the pandemic, as described by Mary. *"I have difficulty hearing people with masks on, I'm continually missing the punchline or the most important part of the communication."*

Difficulty hearing higher pitched voices

Difficulty hearing higher pitched voices is experienced by more than half (55%) of respondents and 26% reported this as one of their top three concerns. Attendees found women's and children's voices especially difficult to hear.

Anna, with hereditary progressive hearing loss beginning in her 20s, described how, *"I had trouble hearing my youngest son, who has a high-pitched voice when we were talking in the car, and he was in the back seat, or when he was calling to me from another room."*

Ron described his difficulties hearing his 13-year-old grandson, *"He speaks very softly and I have a hard time hearing him."*

Difficulty hearing consonants

Half (51%) of respondents reported having difficulty hearing consonants and 18% reported this as a top health concern. For Roz, *"I sometimes mishear certain consonants, which causes my mind to wander off, to consider the words I think I heard."*

Roxana experienced progressive hearing loss at the age of 25 due to an autoimmune disease. She works in the international film industry and said, *"The first challenge I noticed was how some accents started to become very hard to understand. With time, things started to get worse, and what is called a drop in my hearing started to be more noticeable for me."*

Tinnitus

Tinnitus is experienced by 39% of poll respondents and 16% reported this as one of their top three health concerns. Tinnitus was very frequently mentioned during the meeting and in submitted comments. Many people with hearing loss reported that tinnitus was one of their first symptoms of hearing loss, tinnitus is worse without hearing aids and is sometimes so loud that it interferes with sleep. For Anna, *"The first symptom I noticed was tinnitus, or ringing in my ears. Even though it would bother me at night, I still was lucky enough that I could sleep."*

Alissa, who experienced sudden, single-sided hearing loss at age 35 said, *"Tinnitus is masked when I'm wearing my external processor. So, when I remove it in the evening to change the batteries, ... I do notice that it does reappear."*

Christiaan said, *"Due to my tinnitus, I have difficulty sleeping and concentrating. It has negatively affected my sleep, which makes me sleepy and drowsy throughout the day."*

Voice of the Patient Report: *Living with Sensorineural Hearing Loss*

For Alexandre, *“The worst symptom is tinnitus by far. It makes it harder to follow my classes, makes me heavily depressed, causes suicidal ideation and many more. It has essentially turned my life upside down. And as of now, nothing can be done, and it can only get worse if I expose myself further to loud noises. Thus, I am afraid every day of a worsening and have withdrawn from many social activities that people my age engage in (parties, concerts, etc.).”*

Tim said, *“My tinnitus is getting worse as my hearing gets worse. I have no comprehension of what having a quiet moment in my head sounds like.”*

Trouble with balance or vertigo

Vertigo was a health concern reported by 30% of poll respondents and was reported as a top three health concern by 9%. Some attendees described how vertigo preceded hearing loss. Ron described how, *“Every time I would have a severe case of vertigo, it would knock me out for 14, 16, 18 hours. Literally I would be unconscious for a good bit of that time and the time that I was conscious I wasn't able to really do anything. I just was like a wet noodle, just lying there in bed. And each time I would have an episode, I noticed that there was a decline in my hearing.”*

Geri described sound-induced dizziness (Tullio phenomenon) and how, *“If it is a noisy environment, my tinnitus and dizziness rages for several hours afterwards.”*

For some, vertigo came after the hearing loss. Jake reported how, *“Nearly a year after my initial injury, I started noticing I was having balance issues. I started developing vertigo, which turned into vestibular migraines, which have basically taken control of my life.”*

Headaches

While 10% of participants reported experiencing headaches, this was not selected as a top health concern. Jake described vestibular migraines above. Eloise, who was born with hearing loss, described multi-focused migraines. *“I had headaches all my life just trying to hear but I just didn't know what was wrong with me.”*

Other physical health concerns

While 18% of respondents reported that they experienced ‘other’ health concerns, only 5% reported these other health concerns as one of their top three. Panelists, callers and individuals commenting online described a wide range of other physical health concerns that they experienced including hyperacusis (decreased sound tolerance), ear pain or pressure including noxacusis or tonic tensor tympani syndrome, the inability to localize sound, problems hearing and understanding speech in lower frequencies, fluctuating hearing, memory loss and cognitive changes, the inability to hear musical pitch, and diplacusis (when the two ears hear sound differently).

People with hearing loss also experience many psychosocial health concerns such as social isolation, depression and anxiety, verbal communication issues

Social isolation or avoidance

Social isolation or avoidance is a health concern experienced by 61% of respondents and selected as the second most troublesome hearing loss-related health concern by 41% of respondents. Social isolation was a theme that was carried throughout the meeting. Alan described social isolation as the biggest challenge faced by his 97-year-old mother with hearing loss. *“She spends most of her day reading or working on word puzzles and limits her interaction with people she knows she will have a difficult time communicating [with].”*

Munro also experiences social isolation. He said, *“I’m 85 and I have age-related hearing loss. I know people who I have been friends [with] who are pulling away and avoiding me because I can’t hear what they are saying.”*

But this does not just affect elderly people, as Jake, a young man, described. *“I feel isolated from my family and friends. It really feels like I’m sort of watching everyone else live their lives while I’m stuck in place, hoping maybe one day to return to normal life.”*

James, who has genetic progressive hearing loss in both ears beginning at age 8, described how, *“Social interactions are exhausting for me. As my hearing degrades, the cost-benefit of those interactions shift and I find myself opting out more and more, although it doesn’t really feel like a choice. COVID-19 provided relief for me. No more large gatherings, no need to balance my desire to interact with the stress and frustration of not being able to participate fully in the conversation.”*

Mike, who has hearing loss from otosclerosis, agreed and said, *“I’m not sure normal hearing people really understand the stress and anxiety that we experience in social settings.”*

Mood changes: depression or anxiety

Hearing loss extracts a very heavy emotional toll. A third (35%) of poll respondents reported experiencing mood changes such as depression or anxiety, and 7% reported this as a top health concern. Tim described background noise and worsening tinnitus as, *“Feels like a dark cloud hanging over me that just won’t go away, so it’s all I can do to just accept the situation and do the best I can.”*

Linda said, *“Having [tinnitus and hyperacusis] is a monumental challenge because they are externally invisible and often friends and family do not understand the deep depression that can occur.”*

Jess, who has bilateral hearing loss from Alport syndrome, described how, *“Long-term consequences of chronic social isolation due to stigma-enforced communication challenges can*

Voice of the Patient Report: *Living with Sensorineural Hearing Loss*

lead to severe depression, anxiety and the development of mood disorders and/or suicidal ideation."

Along with depression or anxiety, many people with hearing loss experience panic, grief and feelings of loss as they struggle to accept their diagnosis and, in some cases, adapt to what James described as, *"The unrelenting march of a progressive hearing loss."*

Roxana described how for her, *"Hearing loss involves constant loss ... I always say it's like a mourning process. Every time you have a drop, you need to mourn that loss. And for some days, you will feel very sad and shocked because you know it will get worse."*

Katherine, who experienced progressive hearing loss beginning at age 30 said, *"As my hearing worsened over the years, I went through periods of panic and depression. It took me a long time to learn to live with hearing loss."*

When learning that he had permanent hearing loss, Jake described how his doctor, *"Came in the room and apologized to me and told me that there was really nothing he could do for me. I came home, sat at my kitchen table and put my head down and started sobbing. The gravity of the situation completely emotionally overwhelmed me, and I had this realization that my life, as I knew, it was basically over. And this was my new reality."*

Issues communicating verbally

A third (35%) of poll respondents reported having issues communicating verbally and 19% chose this as a top health concern. Several described how they have a hard time communicating when they are shopping, doing errands or travelling. Others described challenges participating in group conversations.

Tim described his verbal communication challenges as a child with hearing loss, *"I started to realize the struggle to communicate with my classmates. My speaking vocabulary was limited, I missed words spoken to me, and I struggled to understand and just keep up. ...You see, for me, it's not just hearing loss, but communication loss. I sounded off because I couldn't speak what I couldn't hear."*

Bill and his wife both experience hearing loss and described, *"Despite the aids, we suffer from a major communications 'gap'. We need to work really hard to understand each other, often still mishearing what we are saying to each other. ... We now need to be in the same room to have conversations—no more shouting down the hall for instance."*

Voice of the Patient Report: *Living with Sensorineural Hearing Loss*

John described how, *"In an attempt to communicate with me more clearly, some people exaggerate their mouth and facial movements. Rather than being helpful, this inhibits my ability to speechread."*

Communication challenges often lead to the misperception that people with hearing loss are less indigent. Tony, born with hereditary SNHL, said, *"What's bothering me the most now, as an adult, is the fact that sometimes because I don't hear all the conversations, it's kind of hard and difficult to ask a question and even answer a question because there have been times when I've answered the wrong question, because the brain didn't put it together... And then I realized people starting to smile, laugh. And so, I kind of like withdraw. ... And so, a little bit later in the conversation, I may ask a question. I'll say, 'Well, what do you think about...?' And somebody will say, 'We already talked about that.' So, it's very embarrassing. And so again, you tend to get more isolated when that kind of happens."*

John added, *"Frequently, I find myself apologizing for not hearing comments, directions and feedback in public settings. Occasionally, people find my misunderstanding humorous. This hurts my feelings."*

Loss of initiative/interest in work/hobbies

A quarter (24%) of poll respondents reported a loss of initiative or interest in their work or hobbies as a result of hearing loss and 6% reported this as a top health concern. Sarah said, *"Meetings with a group, e.g., book club, lectures, plays in auditoriums, are difficult."*

Luke's noxacusis [noise-induced ear pain], hyperacusis and tinnitus, *"Have made it extremely difficult to conduct work, enjoy leisurely activities, socialize, enjoy hobbies from when I had normal hearing ... I have given up several hobbies and pastimes because even with considerable hearing protection present, the activity still aggravates both the pain, hyperacusis and tinnitus too much to continue doing so."*

Other psychosocial health concerns

Panelists, callers and those submitting online comments described additional psychosocial health concerns: hiding hearing loss, stigma, low self-esteem, frustration.

Hiding hearing loss. Meeting attendees described hiding their hearing loss from their families, friends and colleagues by pretending to hear or understand what others said. Some hid hearing loss so that they wouldn't lose their jobs. Some parents described how they tried to hide their hearing loss from their children because they didn't want them to know what burden lay ahead.

Joy, who has Alport syndrome that led to progressive hearing loss said, *"I'm trying so hard not to let my hearing loss be known to [my son], although he knows it, but I don't want him to see it"*

Voice of the Patient Report: *Living with Sensorineural Hearing Loss*

as a burden because if I know that he'll be getting the same hearing loss. So, it adds that extra element of guilt and frustration for both of us."

Stigma. Throughout the meeting participants described hearing-loss related stigma. Roxana described how, *"There is a huge self-stigma with hearing loss. It's not easy when you operate a hundred percent in the hearing world to tell someone you don't hear well. And also, to show your hearing aids."*

Stigma can lead to treatment delays. Julie said, *"The stigma that continues to be associated with hearing loss, especially of adult onset, is a huge barrier. The mental health aspect needs far greater attention and understanding. We have to realize that this unfortunate 'stigma' is related to not getting treatment."*

James said, *"Hearing aids would have benefited me as early as high school and certainly in college, but I couldn't bring myself to take the plunge. I was doing fine, I thought. Hearing aids were for older adults. I get one eventually, but not yet. But like many with hearing loss, I was a terrible judge of what I was missing."*

Low self esteem. Jess described how her mood changes and mental health impacts resulting from the stigma of hearing loss and the deficit-based perspective of the medical system, *"Has led to very significant questions of self-worth. It has contributed very largely to my mental health. I experienced a lot of suicidal ideation, depression, anxiety, chronic PTSD and trauma affiliated with a lot of my experiences from being oppressed just by the system."*

Tim described how he experienced *"Low self esteem issues and isolation. I carried these struggles throughout my school life...but it didn't stop the anxiety and fear of hearing loss-related embarrassment."*

High frustration levels. Throughout the meeting, people with hearing loss described how frustrated they felt when they are unable to hear, unable to communicate clearly, unable to participate, or when things take much longer. Frustration was mentioned 18 times during the meeting and 14 times in the written comments.

People with hearing loss described how hearing loss significantly impacts all aspects of their lives

Meeting participants described the many impacts that hearing loss has on all aspects of their lives, some of which were profound and unexpected. Many described hearing loss as an invisible disability that is often not recognized or acknowledged. Roxana said, *"Hearing loss is called the invisible disability because no one seeing you knows you have hearing loss. ...So, it's complicated for many to understand."*

Voice of the Patient Report: *Living with Sensorineural Hearing Loss*

Darja, who experienced progressive hearing loss beginning at age 22, described how hearing loss is often not recognized even by her professional colleagues in health and social care.

“Unfortunately, many people who are professionals and work with disabled people are not able to understand what [this hearing loss] disability is.”

Zina described how, *“In almost every educational setting, I have been an underrepresented minority and almost always the first person with hearing loss that my colleagues and instructors have ever encountered.”*

Attendees used online polling to select the three most important activities of daily life that they were unable to do or that they struggle with due to hearing loss. The poll results are in **Appendix 5, Q3** and are listed below in descending order. They are followed by the impacts of hearing loss that were not captured in the poll but were described by meeting participants and in the written comments.

Excluded from participating in social events

As Helen Keller said, *“Blindness cuts us off from things, but deafness cuts us off from people.”*

This was confirmed by the polling results, where 88% of poll respondents reported that they are unable, or struggle with participating in social events as a result of hearing loss. This was illustrated by so many of the stories and comments made during the meeting. Brenda, who has hereditary progressive hearing loss beginning at age 19, described the impacts of her hearing loss. *“The result was a severely reduced quality of life, leading to self-isolation, sadness, feeling alone and locked out of life in spite of a very supportive husband and family.”*

Roxana described how, *“My social life started being dominated by my hearing loss. I stopped going out with friends that were hard for me to understand, but at the time you don't realize it.”*

Anna described her challenge, *“I would have a lot of trouble hearing full conversations in a loud restaurant. Socially, loud bars and coffee shops are settings that make casual conversation with friends stressful, even if I'm wearing my hearing aids. This has resulted in some embarrassing misunderstandings and makes what should be a relaxing night out stressful.”*

Barbara Kelley summarized the situation by saying, *“It's relationships with people that make our lives full and hearing loss really cuts you off from people.”* Some of the social impacts are listed below.

Hearing loss impacts friendships. Marcia felt that her hearing loss impacted her friendships. She described the *“Feeling of being left out, not having the joy of following along and always having to listen for key words to know what is going on.”*

Hearing loss impacts the ability to form intimate relationships. Dana described, *“I have been dealing with social isolation because of my hearing loss and probably lost many life*

Voice of the Patient Report: *Living with Sensorineural Hearing Loss*

opportunities because of it, especially where intimate relationships are concerned.” For Tim, “Dating for me was defeating. While guys had stylish, short haircuts, I continued to cover my hearing loss with what I call an ‘outdated seventies’ haircut’ that won’t go away. Not only did I look out of fashion, speaking on the phone was torture. Could I focus well enough to understand? But also, imagined the voice on the other end noting my voice sounds a little funny. Like something is not quite right with me because I’m mispronouncing words. I cannot hide it on the phone, so I just never could let go and just be myself.”

Hearing loss impacts the social lives of other family members including children. Joy described how being the *“Mother of three children ... in a community of hearing people. So, hearing other mothers in social settings for their sports or their school is extremely challenging.”*

Social isolation is particularly hard on younger people. For Jake, *“Being younger, I almost feel like there’s an invisible barrier between me and my social life and people my age. Stuff like going out with your friends, going to noisy restaurants, going to the movies, etc., things that no one even really thinks about have become daily battles.”*

Some reported being purposely excluded or bullied as children. Tim described how, *“I started kindergarten at a regular school. And at first, I thought, how cool was it to show off my hearing thing gadget to my classmates. I felt inferior, and my classmates treated me as such because they didn’t know better. Teasing, mocking, how I talked and bullying was a daily experience for me.”*

COVID-19 has increased social isolation for many people with hearing loss. Masks interfere with speechreading and social distancing means many couldn’t stand close enough to their friends and colleagues to hear. For Mike, *“The pandemic social distancing and the mask wearing has made [social isolation] significantly worse. It would be great to go out to dinner or social settings and be able to hear what the other person or people were saying at the table, or to hear my wife and daughter in the car when we’re driving. Right now that doesn’t occur.”* Ali, agreed, *“Maybe when masks come off, I’ll feel a little better.”*

Conversely, some found that the pandemic offered a social reprieve. Katherine said, *“I am not looking forward to the post-COVID-19 world. My hearing friends can’t wait till we go back to restaurants and dinner parties, but because it’s almost impossible for me to hear speech in noise, that means going back to sitting at a table and smiling because I can’t follow a word being said.”*

Challenges interacting with family members

A total of 57% of respondents reported that they struggled when interacting with family members. Joy described the difficulties in communicating with her son. *“I have a very difficult*

Voice of the Patient Report: *Living with Sensorineural Hearing Loss*

time hearing my son. ... between his speech communication problems and my hearing problems, it's a big fat roadblock for us."

Darja described how she needed to educate her family members about hearing loss. *"I lost my humor. I was completely down and ... my children, my mom, my dad, and they didn't know anything about hearing loss. And then I must explain what is going on, but I was completely down."*

Genevieve and her husband both have hearing loss. *"The words missed in our conversation often lead to total misunderstanding. So far, no divorce! Fifty-five years of marriage helps to keep us together, but hearing loss does not help us to maintain an even keel."*

Unable to attend concerts and events

Almost half, or 43% of respondents reported difficulties in attending concerts or events. Ali described his challenges, *"Just being with friends, going out to dinner or going to a concert or going to a house party, pool party, you name it where there's lots of people around. And it's so difficult to try to have a conversation, even with my hearing aids on or even before I was functional. I was able to do anything I wanted and now it's just, I feel more isolated, struggling."*

Because of hyperacusis, Michael who has noise-induced hearing loss said, *"Had to give up things that I used to enjoy. ... I can't go to concerts anymore, I can't go to movie theaters anymore, even with earplugs because the average concert or movie theater is over 100 decibels these days."*

Wynne said, *"I struggle when I go to public events, theater productions, lectures, etc. Even though it is an Americans with Disabilities Act requirement to have 'assistive listening system', not every venue/location has a system that is working, have knowledgeable staff, or have the AV sound system balance."*

Some people with hearing loss avoid musical events because, like Karen, they experience the *"Loss of ability to enjoy music. It is all distorted."*

Jean described that because of her diplacusis, *"All music sounds off pitch, off. People singing are singing off key. Music I know well is not recognizable to me. ... It makes me dread most forms of entertainment because they are accompanied with music which isn't music."*

Difficulties attending school or work

Only 29% of poll respondents reported hearing-loss related challenges with attending school or work, however the many comments made during the meeting and through the online comments described the profound impacts of hearing loss on education, work and careers.

Voice of the Patient Report: *Living with Sensorineural Hearing Loss*

Employment is much harder for people with hearing loss. Anna described the extra efforts that her hearing loss requires, *“I’m forced to repeatedly ask for my colleagues to repeat themselves and clarify.”*

For Tim, *“I worried for days before speaking in front of people, for fear could I understand if they ask me questions? What do I say in a meeting when my soft-spoken manager is asking me for advice? How in the world do I handle this conference call with people with thick accents? I can’t tell you how exhausted I would feel afterwards.”*

Hearing loss-related fatigue plays a major role in work challenges. Linda, who experienced hearing loss during adulthood said, *“There was a point when literally, if I listened for hours a day, I was so tired I couldn’t do my job. Thankfully, I was within a few months of being implanted so I was able to keep my job. But you have to be able to walk into life, you have to be able to listen six to 10 hours a day without being fatigued. And when you are limited to a couple [of] hours, you have serious limitations on the kinds of things and kinds of relationships you can have.”*

Suzanne, with hearing loss due to Ménière’s disease described how, *“It takes me about three to five times the effort to process people speaking during a meeting. And sometimes I use three layers of captions because [like] captions, not everyone is perfect, but if I triangulate, I get a pretty good idea of what was said and what actions I need to take. But at the end of the day, I’m really tired, and I don’t have a lot left to give.”*

Attending school also requires much more work for people with hearing loss. Tim described his struggles, *“Teachers tried to help, but they simply didn’t understand hearing loss in terms of the emotional and mental toll it took on me. Sitting me in front of the class wasn’t always the answer because hearing loss includes other internal struggles.”*

Zina described her experiences and challenges at medical school. *“I spend significantly more time studying than my classmates since universities rarely teach accessibility, for people with hearing loss. ... It was especially difficult because I was not yet familiar with the terminology to contextualize conversations and fill in the gaps of what I could not hear. I spent countless hours in the lab.”*

People with hearing loss are discriminated from accessing post-secondary education and from gaining employment due to barriers in interview and standardized exams. Speaking on behalf of the HLAA Young Adults Hear group, Zina described that for university students with hearing loss, *“Transitions are particularly challenging, navigating a new environment and*

Voice of the Patient Report: *Living with Sensorineural Hearing Loss*

finding the right resources and accommodations. Socializing and connecting with classmates in freshman dorms can be difficult to hear."

Teri described the difficult experience of a student teacher attending a practicum. *"All the children are wearing masks and she needs to lipread in order to hear them."* Both the remote captioner and sign language interpreters could not hear the children's soft voices, so the student teacher did not pass the course.

Students face other challenges after graduation when finding jobs and transitioning to the workplace. Zina described, *"[The] first step to the interview process is often a phone screen where the recruiter calls the candidate—not accessible for individuals with difficulty hearing on the phone. ... Often, candidates with hearing loss may choose to only disclose their disability once they have received a written offer, fearing that requesting for accommodations during the interview process may put their candidacy in an undesirable light."* Even after obtaining employment, *"Success in the workplace often requires skills related to socializing and networking events in different acoustical environments. Accommodation requests and accessibility are rarely considered for informal events such as happy hours in a bar setting or team dinners."*

Many reported leaving their jobs or retiring early resulting in a loss of income and financial security. Mike said, *"When I was teaching high school, I would come home at night and collapse from the stress and anxiety because of my brain having to work overtime all day, trying to figure out what the students were saying. I tried working with a therapist, but it didn't help. I eventually had to stop teaching."*

Ellie described, *"Early retirement at age 58. My nursing career could have been extended if not for this disability."*

Marcia said, *"I had to quit my teaching career because I wasn't sure the children were reading correctly because I could not hear their pronunciation."*

People with hearing loss are often misperceived as being less capable by employers or colleagues. Attendees at the May 25 meeting represented a range of accomplished professionals, including medical specialists such as anesthesiologists, Ph.D. candidates, decorated war veterans, board members and company directors. Candice said, *"I feel the most dramatic long-term impact is on perceived capabilities in the workplace. ... I find many employers have difficulty looking past an individual's perceived limitations to embrace the many of the skills an individual with a disability can bring."*

People with hearing loss reported being taken advantage of in work situations. Roxana described how, *"I was a young woman entrepreneur, being successful in a male-dominated*

Voice of the Patient Report: *Living with Sensorineural Hearing Loss*

industry, and it was not easy to offset my hearing loss in this environment. ... When I decided to tell a few people about my hearing loss, some people took advantage in negotiations meetings, saying they told me things that they did not."

People with hearing loss often experience a lack of specialized equipment for school or work.

Zina described her difficulty in finding a stethoscope for her medical studies, *"My classmates had a wide range of stethoscopes to select from, ... In contrast, it took me months to find a workable stethoscope. I spent hours doing research online, contacting manufacturers and purchasing different stethoscopes to try. Once I selected a stethoscope, I had to ask an instructor to try my stethoscope and teach me how to use it because it has different features than standard analog stethoscopes."*

The COVID-19 pandemic brought some unanticipated work benefits including online conference platforms, some of which include captions. James described how, *"Teaching on Zoom means I have [a] much easier time of hearing my students"* than in a classroom situation.

Katherine described how, *"Oddly, my best days have also been in the past year. I'm a writer and an advocate for people with hearing loss. Because every meeting was held virtually, I was able to continue working with my committees and groups. I could follow along with captions, either provided by the host or using my phone app. I did the same with social gatherings. I felt more present in the meetings in the past year than ever before in my life."*

For Suzanne, *"When I'm working from home, our team uses MS teams and sometimes WebEx and I can leverage the captions. ... My place of work also equipped me. I was given an accommodation of a dongle so that I can attach, hook up my hearing aid to my work laptop. I'm pretty much on a similar playing field as my colleagues, which is really, really nice."*

Reduced participation in sports/recreational activities

A total of 22% of poll respondents reported that their hearing loss leads to reduced participation in sports/recreational activities. Mike described how all of his leisure pursuits are affected by his hearing loss, *"I have just as much difficulty hearing in a restaurant as I do when I'm outside playing golf or playing tennis."*

Bill said, *"We love our walks in the woods, but we know that we are missing the sounds of birds, rustling leaves and the wind that we recall in our youths."*

For Rand, who lost his hearing after a traumatic accident, *"I cannot drive, shop, go outside, bathe, enjoy music, or even exercise."*

Other impacts of hearing loss

Voice of the Patient Report: *Living with Sensorineural Hearing Loss*

While only 19% of respondents selected “other” important activities of daily life that were impacted due to hearing loss, people with hearing loss described many impacts, below.

Difficulty accessing health care, hospital environments and patient care. Caregivers and patients described the lack of accommodation specifically when speaking to health care providers, and challenges in understanding care recommendations. Katherine accompanied her late husband to his full-day chemotherapy appointments. *“The chemotherapy suite and open floor plan to facilitate communication was a nightmare acoustically. The hospital did not have clear masks. I tried using my smartphone captioning app, but the noise in the room drowned out the speaker so that the captions didn't work. It was intensely frustrating not to be able to hear what nurses and doctors were saying. As my husband's caregiver, I worried that I was missing vital information.”*

Jamie added, *“It is nearly impossible to obtain accommodation in medical settings, no matter how small the accommodation was requested. Nurses continue to call my name from a far-away door, to talk with their backs turned, to get annoyed at me when I explain over and over that I cannot understand. Doctors will not write down what they say. It is just as bad in ENT [Ears, Nose and Throat] offices as at the dentist. Complaining up the chain of command has led to my being assured things will change, but they do not. It is exhausting and maddening and scary how little medical professionals care if I can hear or understand.”*

During the COVID-19 pandemic, many people with hearing loss who work in a health care setting had to change roles or quit their jobs. Anna, an anesthesiologist, described, *“In my job, I have to wear masks in the operating room. Since the pandemic, I wear a mask at all times in the hospital. Masks make it even harder for me to understand conversation as the mask covers up non-verbal expression cues and makes lipreading impossible. I've noticed that the masks also make it very difficult for my older patients to hear me.”*

Ali described how he had to leave his work due to COVID-19. *“I worked in a hospital as a physical therapist. So being in a loud environment, I couldn't function with a mask. I couldn't read people's lips anymore. I couldn't hear with ventilators going, being in patients' rooms. I have to literally stand less than a foot away from somebody to hear them with my right ear, as close to them as I could possibly get. ... I just couldn't function in there anymore.”*

Safety impacts. Some described safety challenges when driving or biking. James said, *“When I'm driving, I can't understand what my kids say if they're sitting behind me. I know it's not safe, but I turn around to see them in hopes of hearing them better. On my bike, I rarely hear cars approaching from behind.”*

Wendy said, *“When walking or driving, I cannot tell from which direction a noise is coming.”*

Voice of the Patient Report: *Living with Sensorineural Hearing Loss*

Challenges with errands and accessing services. Wynne said, *“I struggle with retail and services (banking, vet, groceries, etc.). This is due to reverberation of the physical sound space AND background noise AND people not speaking clearly (then add a mask problem...). I do a lot of faking and try to figure out if the missing information is okay to do without or whether to repeatedly ask for repeat.”*

For Jim, *“One of my biggest frustrations ... is the difficulty using speakers found at drive-up windows found at banks, pharmacies, fast food restaurants, various self-service kiosks, and others. I simply can't understand the speakers and speechreading is not possible.”*

Carole said, *“Obstacles in getting to services (health services, retail services, insurance providers, government services) when using the voice telephone is imposed on me. For really important calls where I must be sure I understand correctly, I need a hearing person with me, and in times of COVID-19, this is often really difficult.”*

Challenges watching TV. This was mentioned often, especially in written comments. Tony said, *“Caption quality is poor, especially with recorded movies. No reason to not be perfect. I avoid people, I do not want to avoid the TV.”*

Louis says, *“Unable to enjoy live TV because captioning is so terrible. It continues to get worse over time. When watching the news for example, captioning is either ‘chunking’ (hesitates then plays catch up so fast you can't read the captioning) or it stops before finishing what was said.”*

Missing out on things that others take for granted. Joy said, *“I can't hear my cats purr, the birds singing, or people coming up behind me when I go hiking.”*

Bill described that with his hearing loss, he and his wife have lost, *“A precious connection to life and, most dearly, our family and friends.”*

People with hearing loss have many worries for the future including increased risk for dementia

Using online polling, people with hearing loss selected their top three worries for the future, which are shown in **Appendix 5, Q4** and ranked below. The many other hearing loss-related worries that were mentioned during the meeting are also included.

Increased risk for dementia

Increased risk for dementia was the top worry for the future, selected by 67% of poll respondents. For Tim, *“I have fears about the future. Dementia and hearing loss seem to show a strong correlation. Will I lose my memory? My mind?”*

Voice of the Patient Report: *Living with Sensorineural Hearing Loss*

Tony said, *“My hearing loss is hereditary, and I see it with my dad. He's already got dementia. I'll probably be there too. ... He wants to die. And I just hope that I don't get to that point.”*

Losing social connections or losing a relationship with a spouse or child

Losing social connections and losing a relationship with a spouse/child were selected as top three worries by 60% and 18% of poll respondents, respectively. For Alexandre, the worry of losing social connections was related to a worry about worsening hearing loss. *“I am afraid every day of a worsening of my hearing loss and have to withdraw from many social activities that people my age engage in parties, concerts, things like that.”*

Anna said, *“I worry about how my hearing loss will impact my ability to communicate with family and friends, as it becomes more severe. I worry that I will have difficulty keeping in touch with my children when they go away to college. I worry that I will have difficulty participating in family reunions and other large, loud family gatherings.”*

For Katherine, *“My biggest concern going forward, now that I'm a widow and living alone, is whether my grown children will trust me with their children. I'm not sure that I would trust me with their children. ... I'm afraid that my hearing loss may make that too risky.”*

Worries selected in online polling

Using the online polling, 43% of poll respondents are worried about **losing the ability to communicate**, 35% of poll respondents worry about **whether they will have the energy to work/live as they want**, 20% of poll respondents identified **not knowing if hearing loss will progress**, 18% of poll respondents worry about **supporting themselves and their families** and 12% of poll respondents worry about **falling**. There were few comments submitted on each of these subjects.

Other worries articulated during the meeting

While only a small percentage (5%) of poll respondents selected “other” worries related to hearing loss, many of these worries were articulated throughout the meeting including worries about **children or grandchildren experiencing hearing loss**, worries about **the lack of assistance** or **the ability to keep up with technology**, worries about a **lack of hearing loss accommodation in the future**, and worries about **increased risks of other diseases**.

James described how, *“I can cope with this for myself, but I can't accept the same situation for my sons and daughter. My two sons, kindergarten and sixth grade, already show signs of loss. The other day my six-year-old said, very matter of factly, that he thinks he needs a hearing aid. I found it very hard to hold it together. ... He really has no idea of the challenges that lie ahead.”*

Voice of the Patient Report: *Living with Sensorineural Hearing Loss*

Darja commented, *“I'm afraid [of] how to keep up with the technology, the technology is exploding now. And it is so hard to follow everything, to connect everything together, to switch from one app to another app and then another app, and that is not so easy.”*

Kate explained how her GJB2 gene defect, *“Also puts me at risk for increased chances of breast cancer, bladder and colorectal cancer, which I just never imagined.”*

Topic 2: Current and Future Treatments

During the second half of the meeting, people with hearing loss described the different technologies, strategies and approaches that they used to manage their hearing loss symptoms and to connect with others. They described how well these approaches worked for them, some of the downsides they experienced as well as suggestions for ways things can be improved in the future.

Different situations need different solutions: people with hearing loss use a combination of technology solutions, medical treatments or medications to address their hearing loss

Meeting participants rely on multiple approaches because different situations need different solutions. Lise, who had hearing loss since birth on her left side and a sudden hearing loss on the right at age 30, explained. *“We use all kinds of tools to help us go from day to day from captioning to assisted listening devices, to speechreading, to alerting devices. We use every tool in the box so that we can communicate with other people.”*

James described all the technologies he uses including his hearing aids which, *“Work well as streaming devices. At work meetings I can couple them to a hearing loop or a remote microphone. My phone will stream calls and podcasts to my ears by Bluetooth. I take Zoom calls on the computer in the same way. But I struggle to understand regular speech. I can't hear my wife when she talks to me from another room. When I watch TV, I rely on closed captioning.”*

Many described how much technology has advanced during their lifetimes and expressed gratitude for the solutions they have. Veronica, who experienced sudden hearing loss at 48, described how cochlear implant technology has evolved. *“My first cochlear implant did not allow me to stream directly into my device. My second, newer model, allows me to have direct connection to cell phones, to the television and any place that has a hearing loop. And it also helps with directional sounds.”*

Lise said, *“I couldn't work and communicate without my hearing aid and my cochlear implant. So that's really the most important tool I have. They are fabulous and I really love them, but they can't do everything.”*

Voice of the Patient Report: *Living with Sensorineural Hearing Loss*

Using the online polling, meeting attendees selected all the technology, therapy or medication solutions that they had used, which are presented below in descending order and in **Appendix 6, Q1**.

Technology—hearing aids and implants

Most of the meeting participants have used technology to address symptoms of hearing loss. Two-thirds (67%) of poll respondents reported having used bilateral hearing aids, 44% used single hearing aids, and 39% used cochlear implants. Only 3% reported using a bone-anchored implant.

Meeting attendees described the transition from a single to bilateral hearing aids or from hearing aids to cochlear implants as their hearing loss progressed. Others reported using a combination of both. After her progressive hearing loss caused her speech recognition to plummet, Brenda described how she transitioned to hearing aids, *“First one analog then two digital, an FM system, a telecoil to use with phone and loop systems and a powerful volume control for the phone.”* Finally, her hearing loss progressed to that point that her hearing aids worked only, *“In quiet environments with no more than two people who enunciated clearly, without accents and no reverberation in the room.”* She described life with a cochlear implant, *“I’m now 18 years post implantation and have upgraded the external processor four times to benefit from advances in technology. I look forward to a completely implanted device.”*

Some described how hearing aids and cochlear implants helped to address not just hearing loss but tinnitus. Alissa described using, *“A cross hearing aid device for people with single-sided loss and also a bone anchored device ... those two devices don’t alleviate the symptoms of tinnitus, so I felt that my only option moving forward was a cochlear implant, which I did pursue. And I now currently have a single-sided implant. Tinnitus is masked when I’m wearing my external processor.”*

Even people with one sided hearing loss experience technology challenges. Kim described how, *“One-sided hearing loss, ... that’s a challenge in itself, particularly when you start to think about devices. So, I’ve used different devices.”*

For Dana, *“Being deaf in one ear and 25% loss in the other, makes life extremely difficult.”*

Medical treatments including counseling, behavior therapy and speech therapy

Almost a third of participants, 31%, reported using counseling or behavior therapy to address hearing loss health concerns and 27% tried speech therapy. Many participants described approaches including cognitive behavioral therapy, tinnitus retraining therapy, psychological support, sound therapy and rehabilitation therapy in more detail.

Voice of the Patient Report: *Living with Sensorineural Hearing Loss*

Zina described how she received speech therapy for more than eight years as a child. *“My therapist would mold my tongue saying, ‘it is not R it is R, curl your tongue’. We would play ‘Go Fish’ together so that we could practice saying words like ‘three.’ My auditory-verbal therapist spoke behind me to rely on my residual hearing in order to discern what was being said. I was frustrated to spend so much time in therapy instead of playing. ... Yet, this experience equipped me with persistence, a strong work ethic and holistic listening. It is ultimately because of speech therapy that I can speak and follow conversations without extensive reliance on lipreading.”*

Michael said, *“I tried tinnitus retraining therapy, which I’m sure other listeners have tried. The thing is, is to a degree, it does help. But when you’ve got tinnitus at 65, 70 decibels loud, it comes to a point where you literally have to learn to get beyond that with your own willpower, because those maskers only work so much, depending on how loud your tinnitus is.”*

Jane said, *“I have tried psychological help and sound therapy, although these treatments have been of limited use.”* She commented that she felt sound therapy worsened her condition.

Don, who lost his hearing in Vietnam due to noise exposure, attributed the rehabilitation courses he received with allowing him to keep his job in the Marine Corps. *“I really got a lot out of it. ... I appealed the ‘recommendation to discharge,’ and I was able to stay in the Marine Corps.”*

Medications

Compared to the use of technology and medical therapy, fewer poll respondents reported using medications to address their hearing loss symptoms, including 28% who used antidepressant or anti-anxiety medications, 14% who used steroids and 8% who tried other medications. Geri described, *“Numerous inner ear injections with gentamicin, initially had oral and inner ear steroid injections when I began this journey. ... I take a daily diuretic which has not had any effect on any of my symptoms.”*

Michael discussed oral medications to reduce and/or manage tinnitus and warned, *“You got to be careful not to get hooked onto Valium and or any of those benzodiazepines that can help lower the volume level of your tinnitus.”*

Not using any recently

Only 5% of meeting participants reported not using any medications, medical treatments or technology to address their hearing loss. Dan explained why he was currently doing nothing medical or technological to manage his hearing loss. *“I’m a concert sound tech. My hearing loss is from 40 years of exposure. Hearing aids would simply up the volume of the failing frequencies, speeding up the deterioration. I can’t really consider anything short of hair regrowth from stem cells or similar. I carefully limit my exposure and bide my time.”*

Voice of the Patient Report: *Living with Sensorineural Hearing Loss*

Luke said, *“There is no treatment. Only using protection (ear plugs) to conserve hearing and slow the degradation from normal wear and reduce the impact of loud noise on already damaged/worn cochleas. ... The reality is slowing the wear (and the increase of tinnitus, hyperacusis setbacks) is the optimal solution. Ear plugs are only so effective; even when worn appropriately, sound insults can still lead to setbacks/further deteriorated hearing.”*

For Lars, whose most significant symptoms are hyperacusis/noxacusis and tinnitus, *“I don't do any treatment. There are no treatments for this at the moment.”*

Most poll respondents use multiple additional approaches to address hearing loss and manage symptoms

Using the online polling, meeting attendees selected all other solutions that they had used in addition to technology, medical treatments and medications. These are shown in **Appendix 6, Q2** and described in descending order below.

Captioning on TV, internet or telephone

Captioning was selected as one of the top approaches used by people with hearing loss; 90% of poll respondents reported using captioning on TV or the internet and 46% reported using a captioned telephone. As Mike explained, *“Captioning is very important for people with hearing aids. I'm using captions on my cellphone when the person I am talking to is difficult to understand or has an accent. I also am using them on video meetings and webinars, and I'm very thankful to the video companies that have made captioning available on their platforms during the pandemic.”*

Captioning is essential for medical students like Zina, who relies on a personal captioner. *“Finding a captioner who can transcribe the highly technical material was challenging, but I'm extremely grateful to have had the same exceptional captioner in undergrad, grad school and now medical school.”*

Captioning doesn't work equally for all, especially for those with reading disabilities. Linda said, *“I've tried about everything that is available to try, but I've also learned sign language because ... I have a reading disability, so captioning is not helpful to me.”*

Assistive listening systems (hearing loop/FM/amplification)

Assistive listening systems are used by 67% of poll respondents. Lise described how a hearing loop works. *“The hearing loop is an assisted listening device used in meeting rooms or large areas, or actually you can have it in small areas too. ... It's linked to a microphone where somebody speaks into, and then it sends a signal to your hearing aid and now cochlear implants too, as long as you have a telecoil in those devices. ... They are a huge help and some people use*

Voice of the Patient Report: *Living with Sensorineural Hearing Loss*

them in their living room to hear the TV, if they're not using Bluetooth or some other way to connect with those."

Russ described how, *"I have looped a television set, which actually works very, very well. It works well in a couple of ways. I can hear quite well and at the same time I can mute the output and my wife or whoever else is there can turn it up to whatever volume they find comfortable."*

Lise described how, *"My cochlear implant and hearing aids needs assistance just like I do,"* which emphasized how these assistive listening systems are necessary to overcome some of the limitations of cochlear implants and hearing aids.

Mike explained, *"[my hearing aids] came with a telecoil and that's helped very much when I'm in a large room with a hearing loop like a church or an auditorium. I've been experimenting with different assistive listening devices, not very successfully, such as microphones or streamers because the hearing aids themselves have limitations when it comes to listening and noise."*

Assistive listening systems also include Bluetooth-linked devices. Mike described how, *"My first success with assistive technology was when I purchased made-for-iPhone hearing aids. I'd been avoiding answering my cellphone for many, many years because of the difficulty hearing people on the cellphone and the hearing aid. The feedback I would get from the hearing aid, whenever the phone pressed up against the ear, was very aggravating. Now I have the phone ring right in my hearing aid using Bluetooth. I tapped the hearing aid and the person's voice goes directly to my ear, without the phone being held. ... Now I have clear phone conversations."* He described downsides, *"Bluetooth does have some limitations. When walking outside the connection periodically disconnects while listening to music and my current hearing aids only allow one Bluetooth connection at a time."*

For John, *"Beaming audio from my smartphone to my hearing aids via Bluetooth has been transformational. It's a far better solution than using headphones. I can more completely comprehend speech and music."*

Modifications/accommodations at home

Modifications or accommodations were used by 59% of poll respondents. Many described different modification and accommodations they used at home, at work/school or in social situations. Lisa described some of the tools that her son uses including, *"A 'Roger' pen that can be set down on a table in restaurant or in a group setting at school to weed out background noise especially in settings with terrible acoustics. [This is a] huge improvement over the big, clunky FM system my son used to use at school, which caused him embarrassment. A BIG help has been his smartwatch that allows a "vibrate" option so that he can finally set his own alarm to wake himself on time for school or work commitments."*

Voice of the Patient Report: *Living with Sensorineural Hearing Loss*

Apps for listening or speech-to-text

Apps for listening or speech are used by 59% of poll respondents. Lise described how, *"I'll use what's appropriate to the situation. Sometimes I'll use a speech-to-text app too. Sometimes that's very helpful."*

Increased nonverbal communication (writing notes/text)

More than half (52%) of poll respondents reported using non-verbal communication. Leo described how, *"My wife must write to me, but no one else wants to take the time to write...it has caused a problem in communication. I get tired and depressed because it is a strain to hear what little I can hear."*

For Brenda, *"In spite of the technology, I cannot hear everyone on the phone. I may ask the caller to text me or use captioning apps if necessary."*

Healthy diet and dietary supplements

Healthy diet was an approach used by 44% of poll respondents and dietary supplements were used by 8%. Participants described how they avoided stimulants and inflammatory foods to reduce tinnitus. For Veronica, *"Eliminating inflammatory foods from my diet helped to alleviate sinus issues and ringing in the ears, also known as tinnitus."*

Joy said, *"I avoid stimulants to try and keep the tinnitus from becoming worse."*

Dana said, *"I can't remember all the so called 'natural' treatments I've tried but I can say that none of them worked. ... So, I gave up on all these products, and diet suggestions etc. that claim to stop tinnitus and improve hearing because THEY DON'T."*

Alerting devices (flashing alarms), increased exercise and stress management techniques

A total of 39% of poll respondents reported using flashing alerts, 38% reported using increased exercise and 23% used stress management techniques to manage hearing loss health effects. Brenda described how after her hearing loss progressed, *"I was forced to communicate visually, caption telephones and entertainment, flashing alerts."*

For stress management, Mary reported success with online and in-person Reiki. With regard to meditation, Kim said, *"Being told to begin yoga, learn meditation, or 'get over it' is not appropriate support for those struggling with hearing loss."*

Other approaches to managing hearing loss symptoms and health concerns

A total of 13% of respondents said that they used “other” approaches to manage hearing loss and described speechreading, physical orientation, relying on others to interpret, protecting remaining hearing, language choice, using American Sign Language (ASL), reading themselves to sleep, hyperbaric chambers, tinnitus maskers and support groups.

Speechreading. For individuals like Katherine, who rely on speechreading, COVID-19 has been devastating. *“I can hear sound, but I can't distinguish speech. I'm a good speech reader, and this supplements my ability to function, but COVID-19 was an enormous setback. Over the past year, I've been unable to understand anything said by someone wearing a mask.”*

Physical orientation. People with hearing loss are forced to be extremely adaptable and position themselves optimally. Gloria, who has chemotherapy-induced hearing loss said, *“In personal conversations, I notice that I have to lean into the person in order to understand or hear what they're saying. If a person turns their back to me and talks, I can't hear it, and I have to ask them to please turn around.”*

Ali usually lipreads, but with COVID-19 and masks, *“I have to literally stand less than a foot away from somebody to hear them with my right ear, as close to them as I could possibly get. It's affected my relationships. I've had to try to be on peoples' left side so I can, they can hear when I'm walking, conversing with people I have to make sure I'm looking directly at them, no loud environments, especially during COVID-19, everybody's wearing a mask.”*

Don said, *“As I grow more knowledgeable about this whole process and more realistic, I recognize that there's a lot I can do, even without a lot of technology, that's going to help myself, by my placement, by my choices, by my self-advocacy and by my continuing to learn about this thing we call hearing loss.”*

Relying on other people to interpret at work and in social situations. Ron said, *“Fortunately for me, my wife is usually around me when I'm around the grandchildren. So, if I have [a] need [for] interpretation, my wife is my interpreter. And the same thing with social outings before COVID-19, I would very rarely go anywhere without my wife, unless it was a business meeting.”*

Protecting remaining hearing with noise canceling headphones or ear plugs. Geri described being unable to tolerate loud noises, music and singing and described how, *“Many times I put ear plugs in my ears”* and said that she, *“Pretty much has to live in a quiet world now.”*

Michael uses earplugs to deal with hyperacusis. *“If I go into a loud public area, even a loud crowded restaurant, I wear my custom earplugs so that I don't pay for sitting in a crowded restaurant, by getting earaches two days later.”*

Voice of the Patient Report: *Living with Sensorineural Hearing Loss*

Language choice. Roxana said, *“Although I’m bilingual, I prefer to speak Spanish because Spanish is a language that has less high-frequency sounds and more syllables on their words. And therefore, it was easier for me to understand than English.”*

American Sign Language. For Jim, *“I have begun to learn ASL but the lack of general ASL use detracts from ASL use as a practical means of communication.”*

Reading themselves to sleep to overcome tinnitus. Joy described how, *“I read myself till I’m nearly asleep because once my hearing aids are out, my tinnitus gets worse. ... So, I read until I’m practically asleep in order to mitigate the sleeplessness that tinnitus can cause.”*

Hyperbaric chambers. Veronica described how she, *“Started a 10-week series of hyperbaric chamber treatments. ...They were one-hour sessions, five days a week in an enclosed chamber with saturated oxygen.”* Unfortunately, *“None of the treatments restored my hearing.”*

Tinnitus masking with white, pink or brown noise. Several callers described this approach including Alissa who uses a white noise app on her phone to match the tone of her tinnitus. *“There’s different types of noise ... white noise and brown noise and pink noise, and you can identify the certain sound that closely resembles it. And by playing it in the background, I am able to even that out for myself.”*

Support groups. Darja said that it was difficult for her to accept her hearing loss but how, *“Finally, when I joined a deaf and hard of hearing organization, of course I found people with hearing loss, and I can share my experience, my fear, my skills and so on. And they help me very much.”*

Acupuncture

Acupuncture was selected as an approach used by 2% of poll respondents. Veronica reported success. *“At one point I had daily acupuncture treatments with over 200 needles placed throughout my body. ... I believe the acupuncture treatments help the most...by alleviating the lingering effects of vertigo, such as loss of balance and fatigue from brain fog.”*

Not doing anything

Only 2% of poll respondents reported doing nothing to manage their hearing loss-related health concerns. Jaime explained, *“I have not done anything to treat [my tinnitus] because I believe that the available treatments are not effective; they are expensive and do not provide real solutions.”*

Voice of the Patient Report: *Living with Sensorineural Hearing Loss*

Most meeting participants reported that their current regimen controls their condition “to a great extent” or “somewhat” but identified many drawbacks

When asked “How well does your current regimen control your condition overall?” 30% of poll respondents reported that their regimen controlled their condition “to a great extent,” 57% reported “somewhat” and 11% reported “very little.” None of the participants reported “not at all” and only 1% responded “not applicable because I’m not using anything.” Poll results are presented in **Appendix 6, Q3**.

When asked to select up to three drawbacks of their current approaches, participants selected “improve my hearing, but not completely” and “high cost or co-pay, not covered by insurance” as the top two choices. Poll results are presented in **Appendix 6, Q4** and are described in descending order below. Some of these drawbacks were illustrated in real time during the meeting.

Improve hearing, but not completely

While hearing devices can amplify sound, they do not improve speech recognition or speech understanding to the extent needed. This was demonstrated by the 75% of meeting attendees who selected this option. Katherine said, *“The most frustrating thing about my hearing loss is that despite having the most sophisticated and most expensive devices, I still can't hear. I know that hearing loss is complicated, but I still think that hearing devices should work better.”* Even with a combination of a cochlear implant and a hearing aid, *“Neither allows me to hear in any way resembling normal hearing. I can hear sound, but I can't distinguish speech.”*

A key treatment challenge lies in addressing the complex, conflicting symptoms of hearing loss. Michael described how, *“The tinnitus, the hyperacusis and the tensor tympani syndrome, they all stem from a common denominator, which is the damage to my auditory system. Because with hyperacusis, you're already sensitive to loud sounds, so the last thing somebody wants to do with hyperacusis is go around with hearing aids, because if loud sound goes off, it's going to be amplified even more by those hearing aids.”* He concluded, *“Even though I have hearing aids that are sitting on my desk, I can't wear them because of my hyperacusis.”*

High cost or co-pay, not covered by insurance

The cost of hearing solutions was selected as a downside by 63% of poll respondents. Participants described how hearing aids or other assistive technologies are not considered a medical necessity by some insurance companies. Kate needs the top technology but feels discriminated against by insurance companies. *“There is a complete lack of any assistance in costs for hearing aids for me, which I will need for the rest of my life. ... every three to five years, ... to keep up with the technology. ... The fact that insurance companies don't consider this a*

Voice of the Patient Report: *Living with Sensorineural Hearing Loss*

medical necessity is very, very difficult for me to understand as a nurse, as a patient, as somebody with a genetic defect. It's like saying you don't need glasses to see, and why do you need teeth in your mouth? You know, hearing is a part of being whole as a person."

Hearing aids must be upgraded regularly. Marcia commented that, *"New technology keeps changing. My being retired and living on a fixed income makes them unaffordable."*

Carla agreed, *"The idea that hearing aids have shelf life is also daunting since it seems we need to spend thousands of dollars every five to seven years on hearing aids."*

Many different devices are required. This was illustrated by the large number of approaches described. Carla continued, *"Supplemental microphones and other hearing assistive devices are needed on top of hearing aids and can be costly as well."*

Others aren't cooperative when I ask for accommodation

A total of 39% of meeting participants identified this as a drawback and described this challenge mostly in work or school contexts and provided many examples. For Katherine, a newspaper editor and writer, *"Although I told my department heads that I had hearing loss, they forgot repeatedly. I was expected to attend daily meetings, to have business discussions on the phone, to understand what someone across the room was saying."*

Anna, an anesthesiologist, described how essential it is for her to, *"Establish rapport with my patients in a very stressful time right before they're about to undergo a procedure or a surgery. ... And while I know that there are clear masks available, my hospital does not use them for patients or staff with hearing loss at this time."*

Mary Ellen described how *"I have trouble getting accommodations for my hearing loss, i.e., church, public programs, etc., this means I cannot appreciate it fully or participate."* Her *"comments to the church leaders do nothing. ...It is frustrating."*

Participants described how they sometimes felt a reluctance to self-advocate. Tim described how much energy it takes to advocate for himself. *"I can advocate, but some days I find that I just don't have the energy. It takes mental and emotional energy to ask people to speak up, talk slower, or show patience with me. And then, they forget."*

Ann explained how she, *"Needed to be taught it was OK to ask people to repeat themselves, speak louder."*

Limited availability or accessibility

Limited availability or accessibility was identified as a drawback by 30% of poll respondents and described throughout the meeting. Suzanne said, *"When I worked in the office, my ability to*

Voice of the Patient Report: *Living with Sensorineural Hearing Loss*

hear depended a lot on which conference room I was sitting in and some conference rooms were fine and some conference rooms were ... an acoustic nightmare."

For Latisha, *"I miss all of what's going on at meetings, unless I'm reading something. But in a regular meeting without the digital platform, I can't hear speech and follow what's happening in meetings so a lot of times I'm excluded."*

Anna said, *"At work during in-person meetings, none of the auditoriums and conference rooms where I work are hearing accessible. In the past, because the main speakers were usually mic'd, I could hear them clearly, but questions from committee members or sidebar conversations were very difficult."*

Even in situations where hearing loss is recognized and accommodated, challenges still exist. This was poignantly illustrated when Phyllis called in during the meeting and was unable to understand the meeting moderator and then apologized unnecessarily for this.

Not very effective

A total of 19% of poll respondents indicated that their hearing loss regimen was not very effective, and throughout the meeting people with hearing loss described the downsides of their devices and the different approaches that they are using.

Even captioning, which many people use, is not perfect as there is a latency between speech and captioning. As an example, Ray described how using cellphones, even with captioning is not effective. *"The captioning is far from accurate. Also, I am bilingual and, even when the captioning phone supports Spanish, it takes so much time to switch, that often the conversation is over before the captioning in Spanish starts."*

This was illustrated during the meeting when Sheila called in and pointed out to the meeting moderator that, *"The captioning isn't keeping up with what you're saying."*

Requires too much effort/time commitment

A total of 10% of poll respondents said that their hearing loss regimen requires too much effort or too much of a time commitment. This includes the effort it makes to learn all the new technologies and the time it takes for hearing aid fittings. James described the process of finding a hearing aid. *"I go to an audiologist's office. She gives me a demo hearing aid and based on a few minutes of trying it out in the office, I choose one that sounds good to me. A few weeks later, I returned to pick up the new hearing aids. Because they can't be reprogrammed remotely, I must return to her office for every adjustment. Some for the better and some not. At some point, I just give up and say, it's fine like this. Not because it actually is fine, but because it*

Voice of the Patient Report: *Living with Sensorineural Hearing Loss*

becomes too hard to justify taking time off of work again for another adjustment and marginal improvement. But I wear these imperfect devices every waking moment."

Robert described how he overcame this issue, "*When I had hearing aids, I bought the programming equipment and programmed them myself. ...that was the best money I ever spent."*

Other drawbacks not included in the polls

While only 10% of poll respondents said that their hearing assistive technologies included other drawbacks, they described additional drawbacks during the meeting and in the written comments. These include environmental/background noise interference, limited space behind the ears, not being able to wear hearing aids in water, hearing device battery and maintenance issues, lack of compatibility between accessories and plastic allergies.

Side effects

Only 3% of poll respondents reported experiencing side effects from their hearing loss solutions. Brenda described that as a result of her cochlear implant surgery, "*A side effect for me was a loss of taste that lasted several months."*

Not applicable/Not using any treatments

Only 3% of poll respondents reported that this question was not applicable as they are not using any treatment.

People with hearing loss prioritized hearing restoration and improved hearing as most important for a possible new drug, device or treatment

The different constellations of hearing loss symptoms experienced by each person requires individualized and personalized treatment approaches. Rather than describing one specific solution, some described what they wanted to achieve. For Zina, "*The ideal hearing loss solution would enable me to effortlessly communicate in group settings, even with poor acoustics and loud background noise. I define effortless communication as the ability to actively engage in conversations without leaning forward, experiencing listening fatigue and asking people to repeat. Such solution, would for example, provide accurate speech recognition in quiet and noisy environments and to produce clear loud sound. The solution should be user friendly, easy to learn and to acquire less than 15 seconds to turn on. Finally, solutions for prelingual hearing loss should enable children to hear in classrooms and facilitate language learning. Most importantly, all technologies should be affordable and covered by health insurance. **Hearing health care should be a right, not a privilege."***

Voice of the Patient Report: *Living with Sensorineural Hearing Loss*

For Joy, *“Future treatments for hearing loss for me and my family would be safe for children, safe for kidneys, safe for water use, easy to use or take, would work for sounds from every angle (think about the safety of driving with hearing impairment and hearing a siren or horn behind you, hiking on trails with mountain bikers who are calling out to you that you can't hear, etc.). Ideally, there would be a way to prevent our genetic hearing loss progression so that all of our future generations wouldn't require hearing aids and/or accommodations.”*

Meeting respondents used the online polling to select their top three most important aspects for a possible new drug or device treatment for hearing loss. The full poll results are described in **Appendix 6, Q5** and the top choices are described in descending order below.

Hearing restoration

Most participants are hoping for hearing restoration, as 80% of poll respondents selected this option as one of their top three. They described how regenerative medicine may potentially address the fundamental cause of hearing loss, regrow the damaged and missing hair cells or correct faulty genes. Ali pointed out that a regenerative approach has the potential to address other hearing loss related challenges and he hoped that regrowing the inner ear hair cells, *“Could reduce the tinnitus, it could improve my balance and some vertigo symptoms.”* Like many others at the meeting, he expressed his wish to be part of a clinical study.

Lise encouraged everyone, *“Let's dream. ... If I could take a pill today and do it and have my hearing restored to what I remember or even better, I would do it in a heartbeat.”* She added, *“Maybe it won't happen real soon, but whenever it happens, I think you'll have a bunch of people ready to do it.”*

Several panelists diagnosed with hereditary hearing loss wished for a gene therapy solution to spare their children and grandchildren from hearing loss. For Brenda, *“Getting at the root cause of the hearing loss... would impact our family tremendously. A great outcome, especially for my grandchildren, would be to stop the progression of the hearing loss, nip it in the bud.”*

Brenda's son, James said, *“Most of all, we need medical treatments that, as my mother said, get at the root cause of genetic hearing loss by suppressing the effects of genes, like DFNA5 or preventing them from being passed on in the first place. That way my grandchildren won't even need to think about hearing or hearing loss.”*

The enthusiasm for hearing restoration was tempered with a reminder from Don. *“Hearing loss is very individualized. Everybody has different expectations. ... I think even with a transplant or hair cell restoration, you still might have some limitations in terms of what you have to work through. So just expect that.”*

Voice of the Patient Report: *Living with Sensorineural Hearing Loss*

Improved hearing

Improved hearing was selected by 72% of respondents as one of the three most important aspects for a possible new drug or device treatment. This would include technology that is attuned to hearing speech, as described by James, *"We need better devices to improve speech intelligibility. ... Essentially, the only reason that I wear hearing aids is to understand what people say, what my wife says, what my kids say, what my students and colleagues at work say. But when I asked my audiologist, which hearing aid will help me hear speech and noise, she says, 'It's hard to say, let's try some and find out.'"*

Mike said, *"My hearing wish list would include ... something—either technology-related or biologics—that would help offset the damage done to my inner ear so that I could hear better in noisy situations. ... I'm not looking for a complete reversal of the damage, but any improvement would make a big difference in mine and my family's lives."*

Improved hearing would include a wider range of sounds including speech in lower frequencies and music. Phyllis, who cannot hear low voices said, *"Nobody has ever been able to come up with what I can do to hear the lower register voices. I have tried many, many different hearing aids, different brands, and they all seem not to be able to help. So, I'm hoping that perhaps at some point, people who are out there who are looking for a challenge in terms of improving the future for others, [could] figure out how to help people ... hear voices in the low register."*

Robert, a former musician with bilateral cochlear implants expressed his wish to hear music better. He explained that most hearing devices are tuned to speech which has a much narrower range than music. *"I can hear quite a bit of music now. I can hear classical music and I can discriminate an oboe from my flute, and cello from a violin, that sort of thing. I don't really hear the bass like I did when I played, I don't hear the real high-pitched sounds and they're distorted."*

Russ would also like improvements in hearing music, *"Hearing aids and cochlear implants have been made specifically ... to focus on speech. And that makes perfect sense to me. But loss of music is a huge problem for a lot of people."*

Improved hearing would include technology that segregates and selects speakers in complex hearing environments. This was described by Ray who said, *"I'm really looking forward to...the ability to use the brain to segregate the speaker in a group that I am listening to. I have heard of some research on using brain waves to feed into the hearing aid or CI to be able to focus on a particular speaker like normal hearing people can."*

Mike agreed, *"I also would like to see manufacturers make continued improvements to make the hearing aids smarter through recognition of certain voices in crowds. And the ability to block out extraneous sounds such as TVs or music in restaurants."*

Voice of the Patient Report: *Living with Sensorineural Hearing Loss*

Improved hearing would include better directional discrimination. For Veronica, *“Short of a cure, I would like to see advances in the directional sounds and distances. Currently, both devices, cochlear and hearing aids have a six-foot hearing range. This means I often have trouble hearing if someone is in another room or across the room. I also cannot distinguish where my phone is or have a sense of direction if the phone rings and it is too far away. I have to constantly place it in the same place so that I can quickly find it when it rings.”*

Decreased background noise

Not surprisingly, decreased background noise was selected by 48% of poll respondents, as this was a challenge mentioned throughout the meeting. Russ explained how, *“Most good top-of-the-line hearing aids have options to decrease unwanted noise, usually focusing on what’s in front of you and suppress what’s on the sides and the back. They work to some degree, but frankly, not terribly well. I think some work on that would be terrific because I noticed that I, as well as many, many other people today have complained about noise.”*

Brenda said, *“The holy grail of hearing device innovation is eliminating background noise and enabling intelligibility. Until this is achieved, devices will fall short and as long as the miniaturization is a goal, it’s unlikely to happen.”*

Slowing or stopping hearing loss

A total of 41% of poll respondents chose slowing or stopping hearing loss as one of their top three choices. Sheila stated that, *“Each year my hearing gets worse. I need to find some ways to keep my hearing from getting worse. Even restore a little bit of it.”*

This was also mentioned by James who said, *“Looking ahead, we need medical treatments to limit the extent of genetic hearing loss, so my children’s loss does not progress as far as my own has. I want them to participate fully in their social and professional lives in ways that my mother and I have not been able to.”*

Helping with tinnitus

A treatment to help reduce tinnitus was a top choice of 25% of poll respondents. Joy said, *“Any treatments that would actually eliminate my tinnitus would be a huge boom because sitting in a quiet room alone and still having a buzzing constantly in your head all day long can be quite stressful.”*

Improving balance

A total of 7% of poll respondents would like a treatment to improve balance.

Other aspects of hearing loss important for a possible new drug or device treatment

Only 6% of poll respondents selected “other,” however they described many options which fell into broad categories of technology improvements, education/information and support and advocacy. These are as described below.

Hearing aids that can be programmed by the wearer or telehealth options for hearing aid adjustments. Michael stated that, *“Hearing aids that could be programmed by the wearer would offer considerable advantage. It’s very difficult to describe to my audiologist what I hear and what settings seem to help speech comprehension.”*

James wished, *“In the very short term, telehealth options for hearing aid adjustments would provide some logistical benefit.”*

Reduced captioning latency. Several attendees requested reductions in the delay between spoken words and the appearance of the caption. Russ explained, *“Focusing on two things simultaneously, for me anyway, is a huge problem. ... If I can hear sound, I can process it much quicker and much better than I can the written word. But the problem is, with latency, I hear things and I look at the captions and what I’m seeing in the captions is not what I’m hearing.”*

Anna agreed, *“Closed captioning that’s not accurate, makes it more difficult for me to follow the discussion, as I’m trying to read and listen at the same time and reconcile the two inputs to make sense of what’s being discussed.”*

Waterproof hearing devices. Several people wished for hearing aids that could be used when swimming or diving. Joy is a swimmer and a swim mom who said, *“It would be so amazing to have a device that I could wear in water and still feel like I could hear and be a part of conversations when we’re in those activities.”*

Lisa agreed, *“A hearing device that allows for hearing in the water would be amazing. ... Would love for people living with hearing loss to enjoy the experience of swimming in the water, lakes and or in pools.”*

More education and information sharing about available options including consumer reports and fatigue ratings on hearing devices. Don said, *“We need to have more consumer-based education as to not only what’s out there, but what is out there that’s reliable and it works for many people based on the severity of their loss.”* To illustrate the value of this, he described how over-the-counter amplification devices, *“Would probably work for somebody with mild to moderate hearing loss, but they’re not going to touch somebody with a severe or profound loss.”*

Linda, a scientist with adult hearing loss, wants standardized fatigue rating scales for devices. *“It’s really important that people understand what they’re buying before they buy it. When I try*

Voice of the Patient Report: *Living with Sensorineural Hearing Loss*

to choose a cochlear implant, having some information about how tired I'm going to be, as I walk into life, it's really important."

Don also emphasized the importance for people with hearing loss to educate each other. *"It takes a lot of hard work, it takes a lot of knowledge, it takes a lot of going back to your peer group and sharing that knowledge so that they would know what your experiences are. And I really think that this education, self-advocacy, they're the keys."*

More recognition and support for hearing loss-related emotional challenges, as well as patience and kindness for those with hearing loss. Many people with hearing loss mentioned the emotional challenges associated with diagnosis and hearing loss progression. They asked for support and counseling, not only for those with hearing loss but for their family members as well. Mike said, *"I would also like to see more attention and solutions to address the emotional challenges that we face. ... I'm keeping a positive attitude as I look forward to improvements for those of us with hearing loss in the near future."*

Lynne spoke on behalf of her husband with hearing loss who, *"Feels a need for a person or group where he can discuss and learn how he can deal with his difficulties and frustrations."*

Michael recommended, *"Audiology clinics need to provide counseling for both the person with hearing loss and the normal hearing spouse or other close family members or friends. Hearing loss requires learning coping strategies that both parties need to learn."*

Dave requested more understanding and cooperation for those with hearing loss.

Greater awareness of hearing loss overall and more training for medical professionals. Jim said, *"I wish there was a better understanding of hearing loss by the general public. When I tell people that I can't understand them they just continue to talk, and I still can't understand. Some of the worst offenders are in the medical community."*

Tim recommended, *"More development with assistive hearing devices and training in the medical and hospital setting,"* with the goal for medical professionals to, *"Have the training and understanding to effectively communicate with their hearing loss patient."*

Lawrence expanded on this and suggested teaching physicians to directly face their patients, to speak clearly, to ask questions about the patient's hearing history or family history of hearing loss and to ask the patient to repeat things to make sure they comprehend.

Voice of the Patient Report: *Living with Sensorineural Hearing Loss*

More widespread early testing, diagnosis and treatment of hearing loss. Sara asked, *“What are we doing to improve hearing loss treatment equity? What can we do to help more people get tested and treated?”*

Eloise said, *“We don't need to be waiting at 50 years of age for people to discover that they're hard of hearing.”*

Joy described how she waited a long time before treatment and encouraged others not to do the same. *“One of the lessons that I learned along the way, was that I try to really impress upon others with Alport syndrome, is not to wait on the hearing aid.”*

Greater recognition of hearing aids as a medical necessity, and reimbursement by more insurance companies and Medicare. Kate stated, *“I would like to understand when the insurance companies are going to get on board and help people like me. It's very, very discouraging.”*

Other recommendations included a fully implanted cochlear implant, more hearing loops in the community for large-area listening with telecoils and more research into the etiology of hearing loss.”

Incorporating Patient Input into a Benefit-Risk Assessment Framework

The FDA uses a Benefit-Risk Assessment Framework which includes the following decision factors: analysis of condition, current treatment options, benefit, risk and risk management. The Framework provides an important context for drug regulatory decision-making, including valuable information for weighing the specific benefits and risks of a particular medical product under review.

Table 1 serves as the proposed introductory framework for the Analysis of Condition and Current Treatment Option rows, which could be adapted and incorporated in the FDA's Benefit-Risk Assessment, help enable a comprehensive understanding of this unique condition for key reviewers in the FDA Centers and Divisions that would be reviewing new treatments for SNHL. The data resulting from this meeting might also help inform the development of clinically meaningful endpoints for current and future clinical trials and encourage additional manufacturers to investigate this condition. The information presented captures the perspectives of people with hearing loss presented at the May 25, 2021, meeting, as well as polling results and comments submitted before, during and after the meeting through the online portal. Note that the information in this sample framework is likely to evolve over time.

Table 1 speaks to the challenges that people with hearing loss face. Hearing loss is an invisible disability and there are so many experiences and environments that are not accessible or

Voice of the Patient Report: *Living with Sensorineural Hearing Loss*

accommodated for those with hearing loss. In addition, hearing loss is not recognized as a medical necessity for reimbursement by insurance companies. Hearing health is critical to living a healthy vital life, yet there is a vast treatment need for ways to improve or even restore hearing for people with SNHL. As James emphasized, *“My family needs better hearing solutions so that we can live our best lives.”*

Voice of the Patient Report: *Living with Sensorineural Hearing Loss*

TABLE 1 Sensorineural Hearing Loss Benefit Risk Table

	EVIDENCE AND UNCERTAINTIES	CONCLUSIONS AND REASONS
ANALYSIS OF CONDITION	<p>Sensorineural hearing loss (SNHL) results from damage to the hair cells or nerve fibers of the inner ear that convert sound into electrical impulses.</p> <p>SNHL encompasses noise-induced and age-related hearing loss, as well as hearing loss caused by ototoxic medications, some viruses, and idiopathic sudden hearing loss. SNHL can be heritable and may present as part of other disorders such as Alport syndrome or Ménière’s disease.</p> <p>SNHL results in decreased sound sensitivity and distortion in sound encoding resulting in a garbled sound signal. People with SNHL have problems distinguishing or understanding speech.</p>	<p>SNHL includes a very complex and interfering combination of physical and psychosocial health concerns.</p> <p>SNHL is a permanent and degenerative condition that can often lead to severe hearing loss.</p> <p>People with hearing loss described many worries for the future. The link between hearing loss and dementia is a particular cause for concern for people with hearing loss.</p>
IMPACT ON ACTIVITIES OF DAILY LIVING	<p>Hearing loss profoundly impacts all activities of daily living. Hearing loss leads to social exclusion and diminished quality of life.</p> <p>Hearing loss is heavily stigmatized. Many hide their hearing loss, are intentionally excluded and misperceived as less intelligent. Many described feelings of denial or shame, which made them reluctant to seek treatment.</p>	<p>SNHL is an invisible disability.</p> <p>Hearing loss accommodation is not recognized as essential, and many environments are inaccessible including hospitals, airports, schools and workplaces.</p> <p>Hearing aids and other assistive technologies are not considered medical necessities.</p>
CURRENT TREATMENT OPTIONS	<p>Addressing the complex, conflicting symptoms of hearing loss is a challenge. SNHL symptom management strategies involve an integrated and personalized approach, including a combination of technology solutions, medical treatments and medication as well as multiple other approaches and strategies.</p> <p>Very few individuals with SNHL report not using any treatment approaches.</p>	<p>Existing SNHL treatments are not curative or disease modifying and cannot reverse or repair the damage already done.</p> <p>Current regimens improve hearing, but not adequately, and often have a high cost or copay that is not covered by insurance.</p>
PROSPECTS FOR FUTURE	<p>People with hearing loss are hoping for hearing restoration, improved hearing, decreased background noise and slowing and stopping hearing loss.</p> <p><i>See the “Voice of the Patient” report for a more detailed narrative.</i></p>	<p>People with SNHL are hopeful for new options to restore or improve their hearing and many expressed a wish to participate in clinical trials.</p>

Voice of the Patient Report: *Living with Sensorineural Hearing Loss*

Conclusion

The May 25, 2021, SNHL EL-PFDD helped advance the HLAA mission to address the unique needs and challenges of individuals with SNHL, by bringing together diverse members from the stakeholder community. This meeting provided an opportunity to hear directly from people and family members about their experiences of living with hearing loss, the impact of hearing loss on their activities and daily lives, and their worries related to their hearing loss. Meeting participants described the different medications, technologies, strategies and adaptations that they had used to address their hearing loss. They identified unmet needs and shared their preferences and perspectives for future technologies and therapies directly with the FDA, academic investigators, industry partners, medical product developers and representatives from government agencies colleagues.

The participants at this EL-PFDD clearly demonstrated that hearing health is critical to living a healthy vital life. At several points during the meeting, people described how much they valued the experience of sound—no matter how imperfect or incomplete. Mike described how the day that packing from his stapedectomy surgery (to replace damaged bone from otosclerosis with titanium replacement) was removed, *“I could hear again with moderate hearing loss, instead of being deaf. It was like having a switch flip from off to on in my brain.”*

After learning how to use an accessible stethoscope Zina said, *“I became teary when I've heard the heartbeat with the stethoscope for the first time. It was one of the most beautiful sounds I have heard.”*

Kate described, *“When I went into those hearing aids for the first time, I realized that I'm not sure when I stopped hearing the birds chirp...I felt extremely grateful to have this ability to hear things that I didn't realize I wasn't hearing.”*

Several months after receiving her cochlear implant, Brenda said, *“My brain identified sounds it hadn't heard for years and returned me to the world around me. Birds, neighborhood dogs, microwave beeps, some music and movies.”*

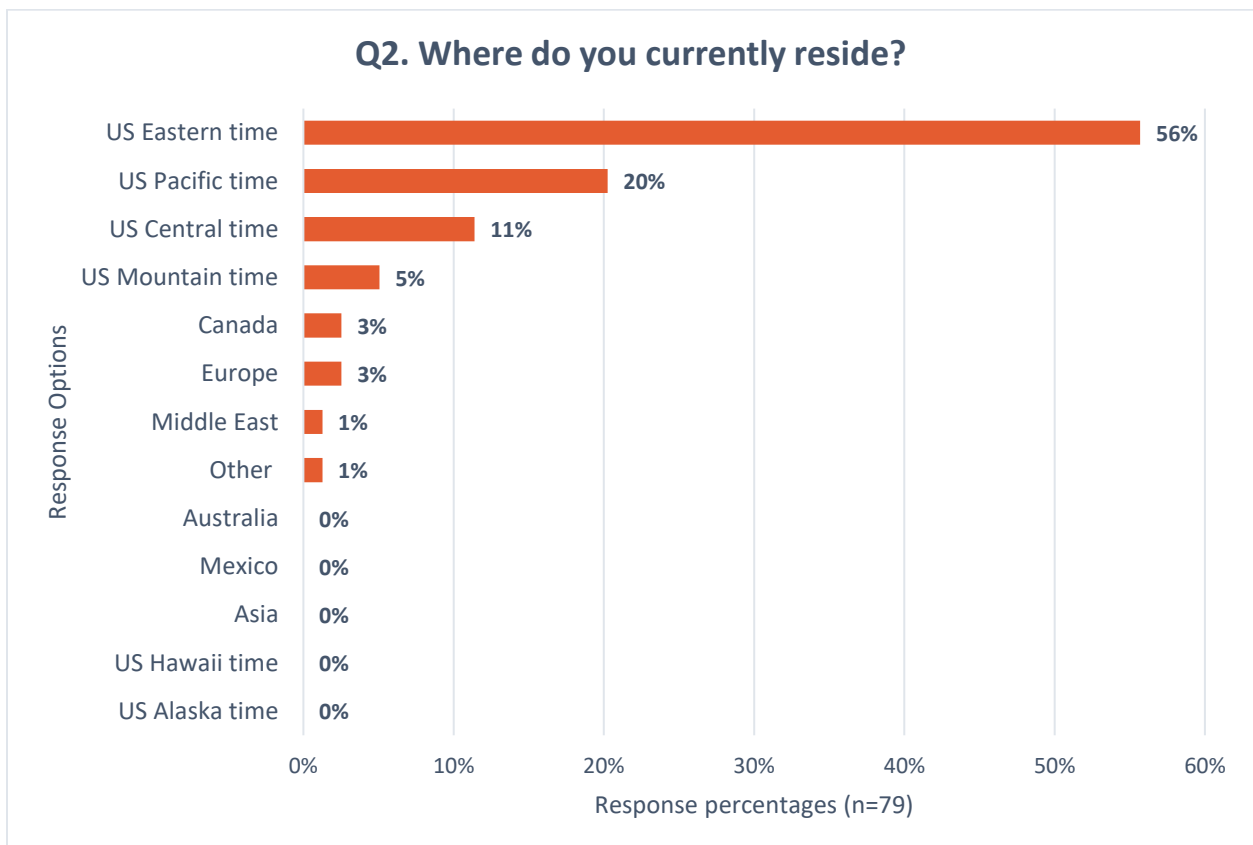
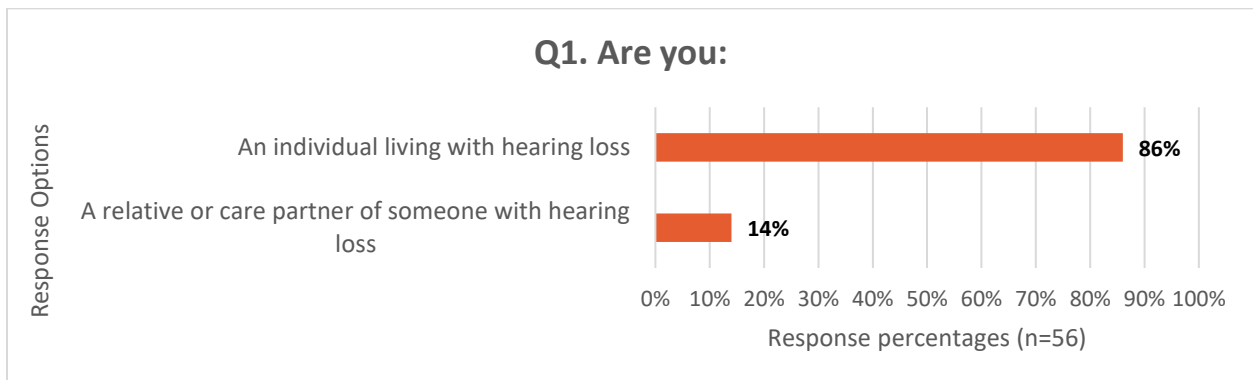
HLAA is grateful for this opportunity to share our community's voices through this “Voice of the Patient” report and, in turn, educate and inspire others to find more and improved solutions to improve the health and quality of life for those living with hearing loss. We believe that the outcomes of this meeting can change the future for patients living with hearing loss.

Voice of the Patient Report: *Living with Sensorineural Hearing Loss*

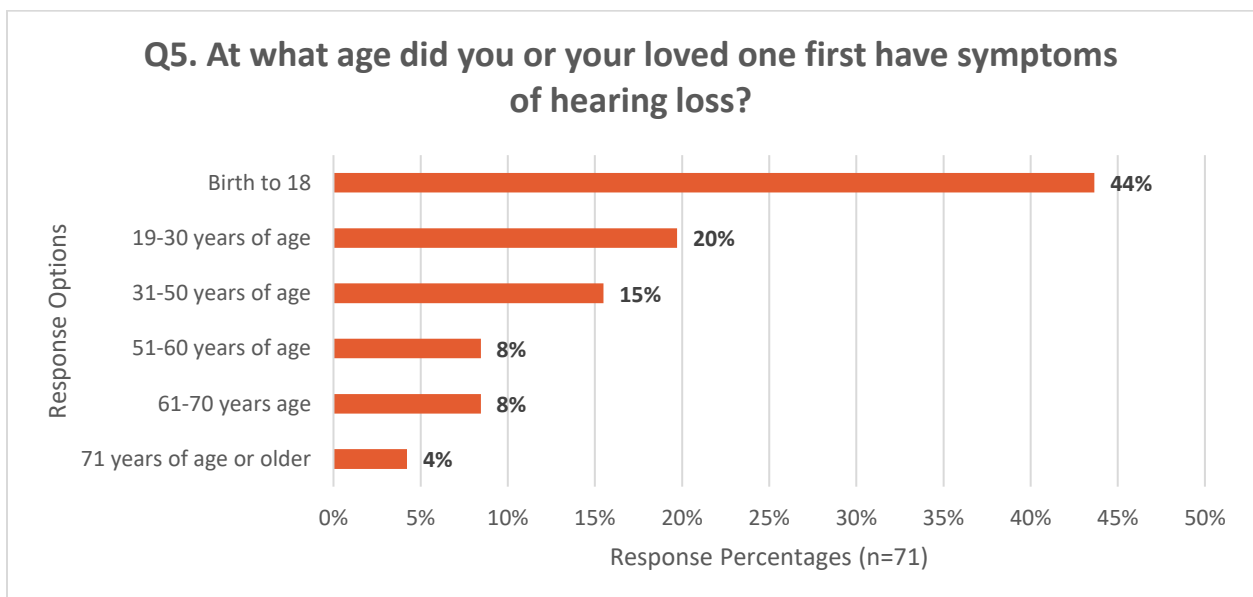
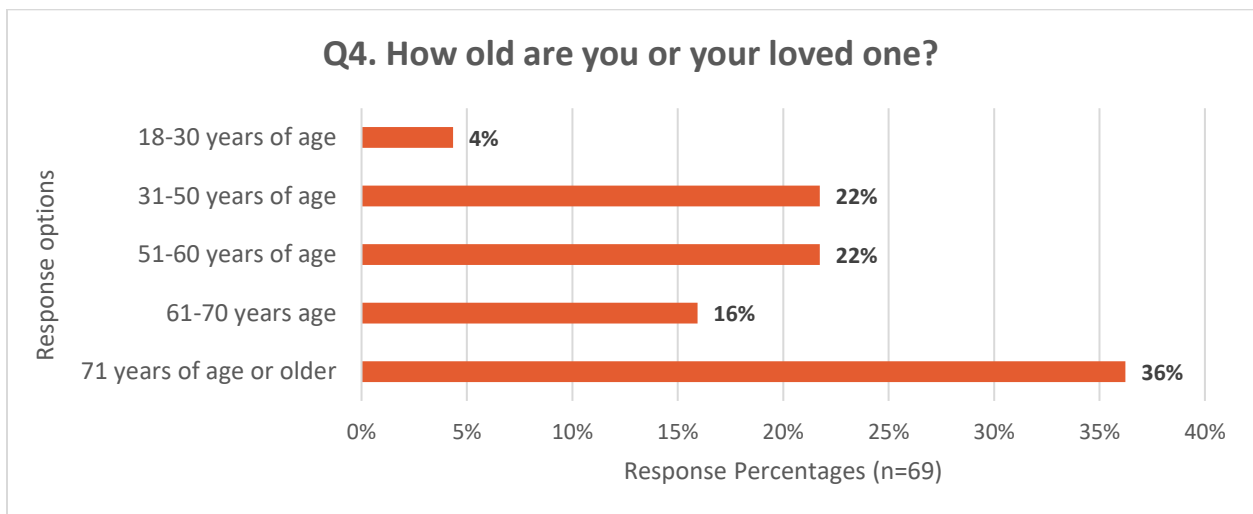
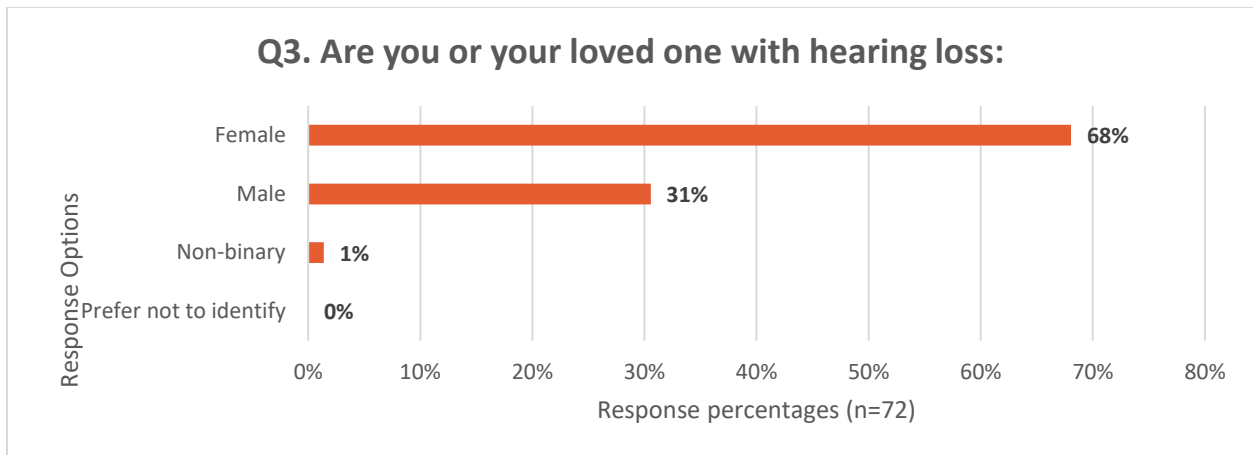
Appendix 1: Meeting Demographics

These graphs include those attendees who chose to participate in online voting. The number of patients who responded to each polling question is shown below the X axis (n=x).

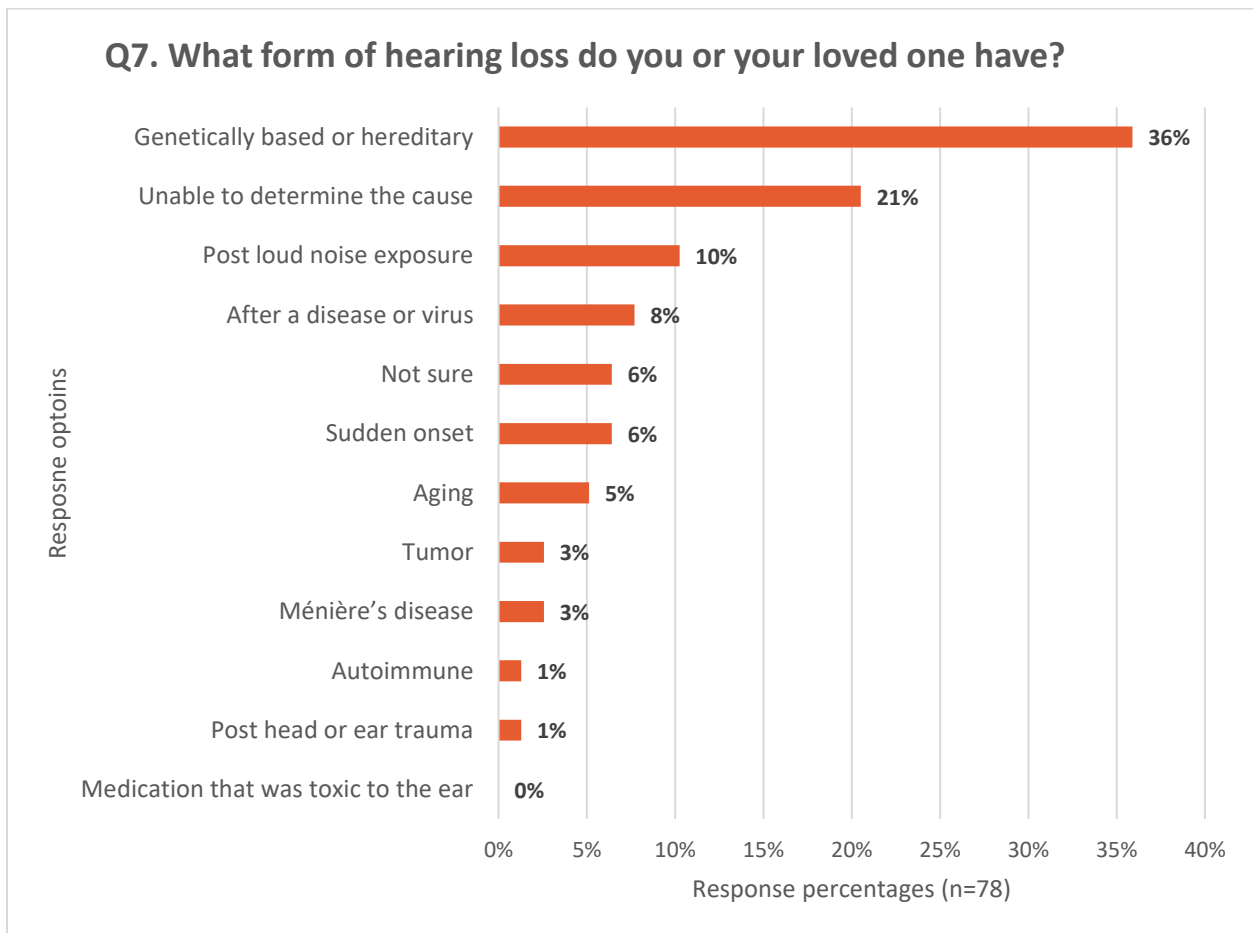
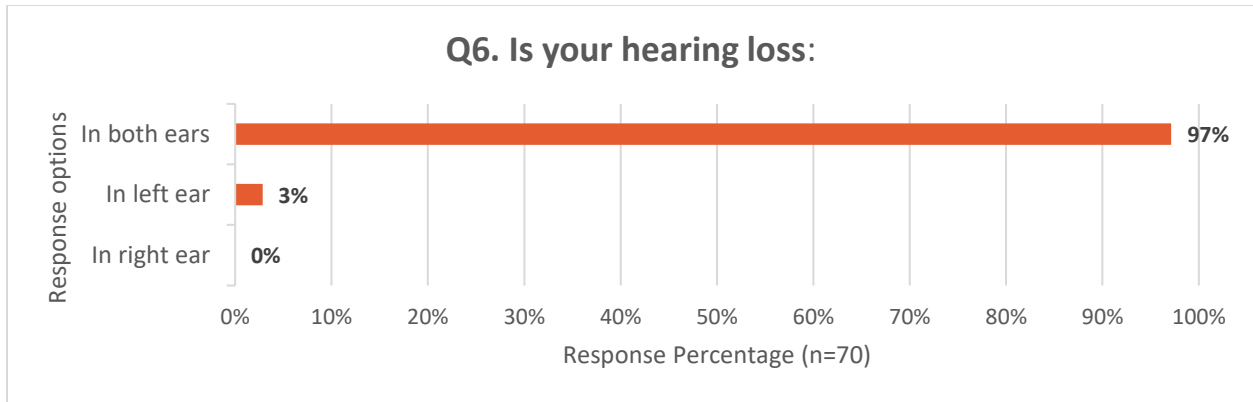
While the response rate data for these polling questions is not considered scientific data, it provides a snapshot of who participated in the EL-PFDD meeting and is intended to complement the live and pre-recorded comments throughout the meeting.



Voice of the Patient Report: *Living with Sensorineural Hearing Loss*



Voice of the Patient Report: *Living with Sensorineural Hearing Loss*



Voice of the Patient Report: *Living with Sensorineural Hearing Loss*

Appendix 2: Meeting Agenda

Hearing Loss Association of America
Externally-Led Patient-Focused Drug Development (PFDD) Meeting
on Sensorineural Hearing Loss

May 25, 2021

10 a.m.-3 p.m. U.S. Eastern Time

www.hearingloss.org/programs-events/patient-focused-drug-development-meeting/

- 10 a.m. **Welcome**
Barbara Kelley, Executive Director, Hearing Loss Association of America
- 10:05 a.m. **Opening Remarks**
Gavin H. Imperato, M.D., Ph.D.
Medical Officer, Office of Tissues and Advanced Therapies
Center for Biologics Evaluation and Research, U.S. Food and Drug Administration
- 10:15 a.m. **Scientific and Medical Overview of Hearing Loss**
Frank Lin, M.D., Ph.D.
Director, Cochlear Center for Hearing and Public Health
Professor of Otolaryngology - Head and Neck Surgery
Johns Hopkins Bloomberg School of Public Health
- 10:30 a.m. **Introduction and Meeting Overview**
James Valentine, J.D., M.H.S., Meeting Moderator
- 10:35 a.m. **Audience Demographic Polling**
- 10:40 a.m. **Session 1: Symptoms and Daily Impacts of Living with Sensorineural Hearing Loss**
Panelist Perspectives
Audience Remote Polling
Moderated Audience Discussion
- 12:25 p.m. **Lunch/Refreshment Break**
- 12:55 p.m. **Introduction to Afternoon Session**
James Valentine
- 1 p.m. **Session 2: Current and Future Treatment for Sensorineural Hearing Loss**
Panelist Perspectives
Audience Remote Polling
Moderated Audience Discussion
- 2:45 p.m. **Summary**
Larry Bauer, R.N., M.A., Hyman, Phelps, & McNamara, P.C.
- 2:55 p.m. **Next Steps and Closing Remarks**
Barbara Kelley
- 3 p.m. **Adjourn**

Appendix 3: Meeting Discussion Questions

Session 1: Symptoms and Daily Impacts of Living with Sensorineural Hearing Loss

Of all the symptoms of hearing loss, which 1-3 symptoms have the most significant impact on your or your loved one's life?

1. How does hearing loss affect you or your loved one on best and on worst days? Describe your best days and your worst days.
2. How have your or your loved one's symptoms changed over time? How has the ability to cope with the symptoms changed over time?
3. Are there specific activities that are important to you or your loved one that you or they cannot do at all or as fully as you or they would like because of hearing loss?
4. What do you fear the most as you or your loved one gets older? What worries you most about your or your loved one's condition?

Session 2: Current and Future Treatment for Sensorineural Hearing Loss

1. What are you currently doing to manage your or your loved one's hearing loss symptoms?
2. How well do these treatments treat the most significant symptoms of hearing loss?
3. What are the most significant downsides to your or your loved one's current treatments and how do they affect daily life?
4. Short of a complete cure, what specific things would you look for in an ideal treatment for hearing loss? When considering a new treatment, what factors are important to you?

Discussion questions to guide online comments (posted on website after event)

1. Of all the symptoms that you experience (or your loved one experiences) because of hearing loss, what are the 1-3 symptoms that have the most significant impact on quality of life?
2. How do these symptoms impact specific activities that are important to you?
3. What are you doing (or what is your loved one doing) to help treat hearing loss, and how well are those treatments working?
4. What are the most significant downsides to those treatments?

Voice of the Patient Report: *Living with Sensorineural Hearing Loss*

Appendix 4: Panel Participants and Callers

Session 1: Symptoms and Daily Impacts of Living with Sensorineural Hearing Loss

Zoom Panel

Roxana, progressive hearing loss caused by an autoimmune disease at age 25

Tim, moderate bilateral hearing loss from birth

Katherine, progressive hearing loss beginning at age 30

Anna, hereditary progressive hearing loss beginning in her 20s

Jake, young adult, sudden hearing loss due to loud noise exposure at age 18

Zoom Discussion Starters

Latisha, born with hereditary SNHL

Tony, born with hereditary SNHL

Kim, gradual single-sided hearing loss due to Ménière's disease beginning at age 40

Ron, gradual hearing loss due to Ménière's disease and/or acoustical trauma

Darja, progressive hearing loss beginning at age 22, unknown cause

Callers Joy, Alport syndrome that led to progressive hearing loss

Phyllis, cannot hear speech in lower frequencies, such as men's voices

Kate, hearing loss due to the GJB2 gene defect

Ali, viral-induced hearing loss

Suzanne, hearing loss due to Ménière's disease

Eloise, born with hearing loss

Session 2: Current and Future Treatment for Sensorineural Hearing Loss

Zoom Panel

Zina, diagnosed with hearing loss at the age of 3 1/2 years

Brenda, hereditary progressive hearing loss beginning at age 19

Mike, hearing loss from otosclerosis, gradual then sudden, starting at age 42

Veronica, sudden hearing loss at age 48

James, genetic progressive hearing loss in both ears beginning at age 8

Zoom Discussion Starters

Lise, hearing loss since birth on left, and sudden hearing loss on right, age 30 onset

Russ, progressive hearing loss for the last 25 years

Don, lost hearing in Vietnam due to noise exposure

Alissa, sudden single-sided hearing loss at age 35

Gloria, hearing loss as a result of chemotherapy

Callers

Jess, bilateral hearing loss from Alport syndrome

Joy, Alport syndrome that led to progressive hearing loss

Michael, noise-induced hearing loss

Linda, hearing loss during adulthood

Ali, viral-induced hearing loss

Sheila, virus-induced hearing loss

Robert, former musician with hearing loss

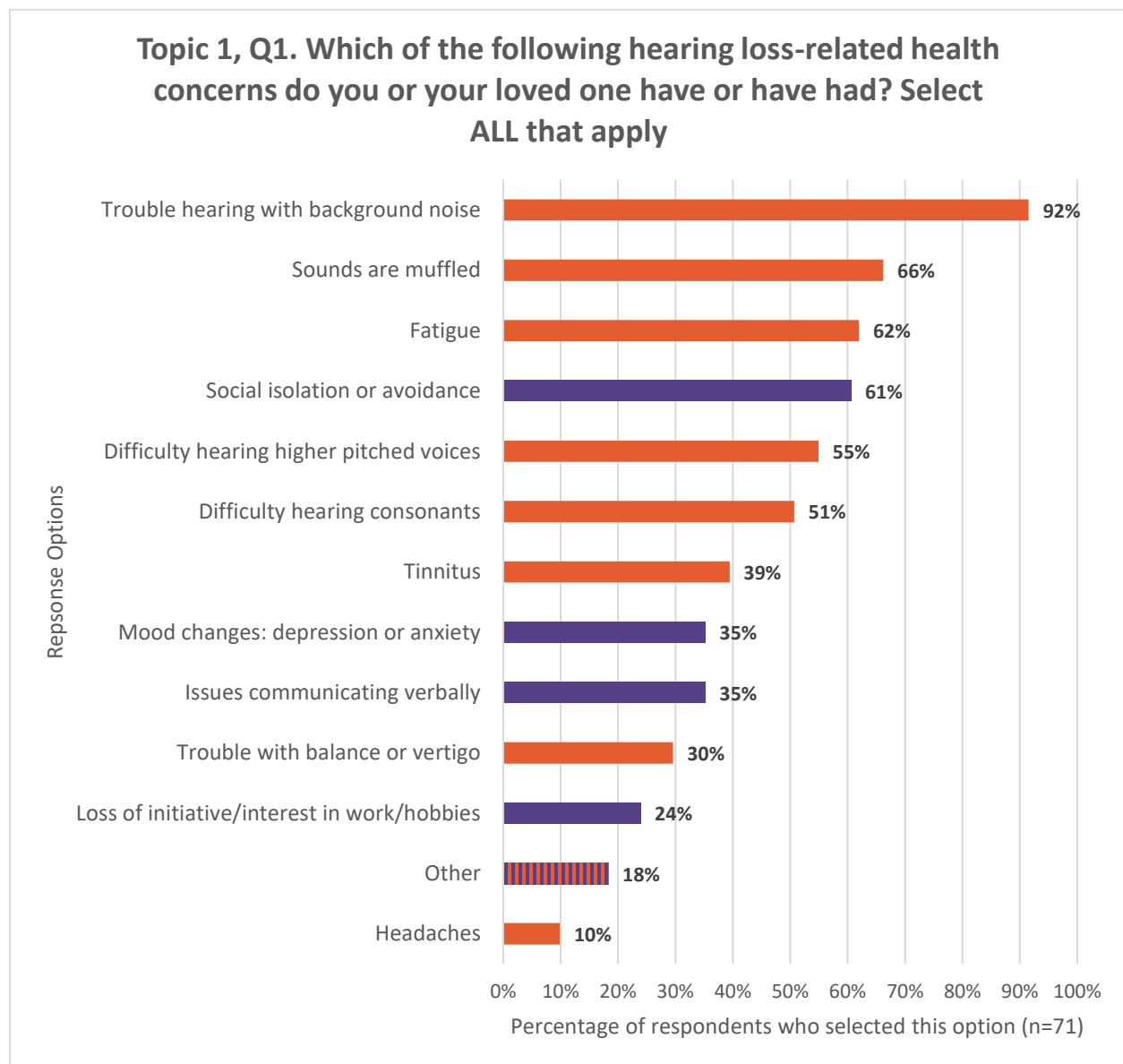
Voice of the Patient Report: *Living with Sensorineural Hearing Loss*

Appendix 5: Topic 1 Polling Results. Symptoms and Daily Impacts of Sensorineural Hearing Loss

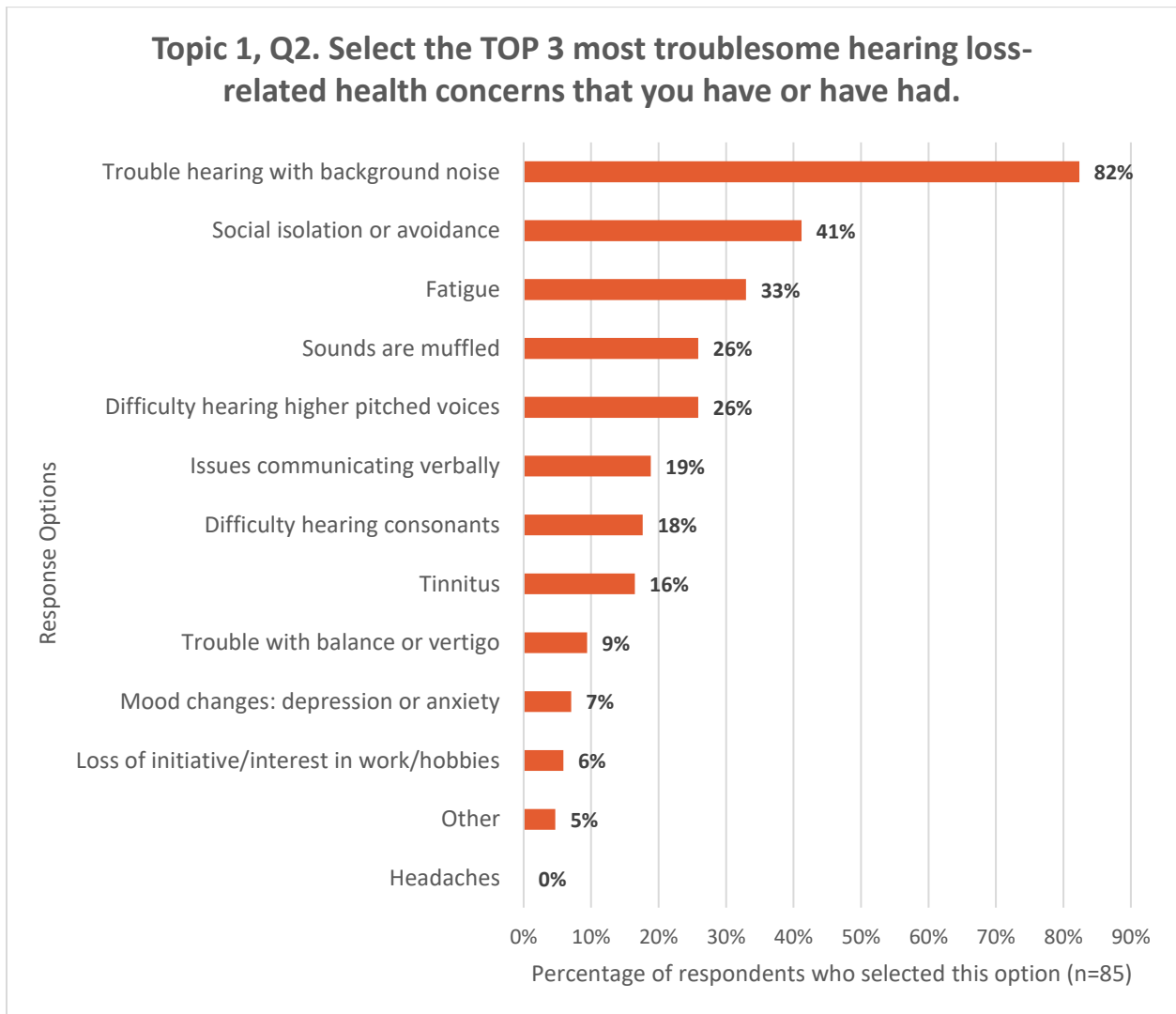
These graphs include those attendees who chose to participate in online voting. The number of patients who responded to each polling question is shown below the X axis (n=x).

While the response rate data for these polling questions is not considered scientific data, it provides a snapshot of who participated in the EL-PFDD meeting and is intended to complement the live and pre-recorded comments throughout the meeting.

For this question, responses were divided into two categories of hearing loss-related health concerns: physical, represented by orange bars and psychosocial, represented by purple bars.

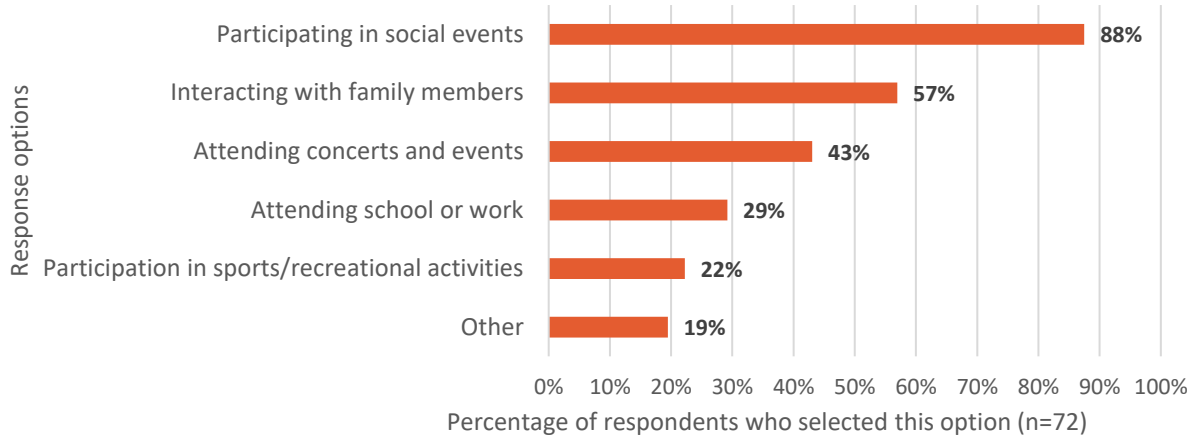


Voice of the Patient Report: *Living with Sensorineural Hearing Loss*

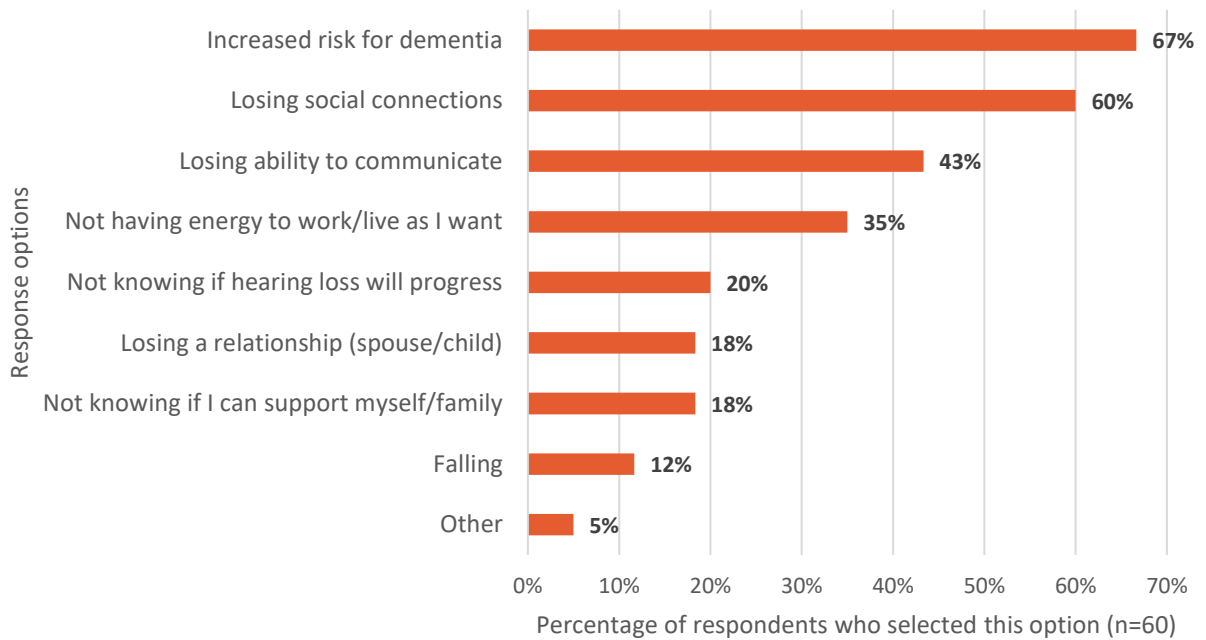


Voice of the Patient Report: *Living with Sensorineural Hearing Loss*

Topic 1, Q3. What specific activities of daily life are most important to you that you are NOT able to do or you struggle with due to hearing loss? Select TOP 3



Topic 1, Q4. What worries you most about you or your loved one's condition in the future? Select TOP 3

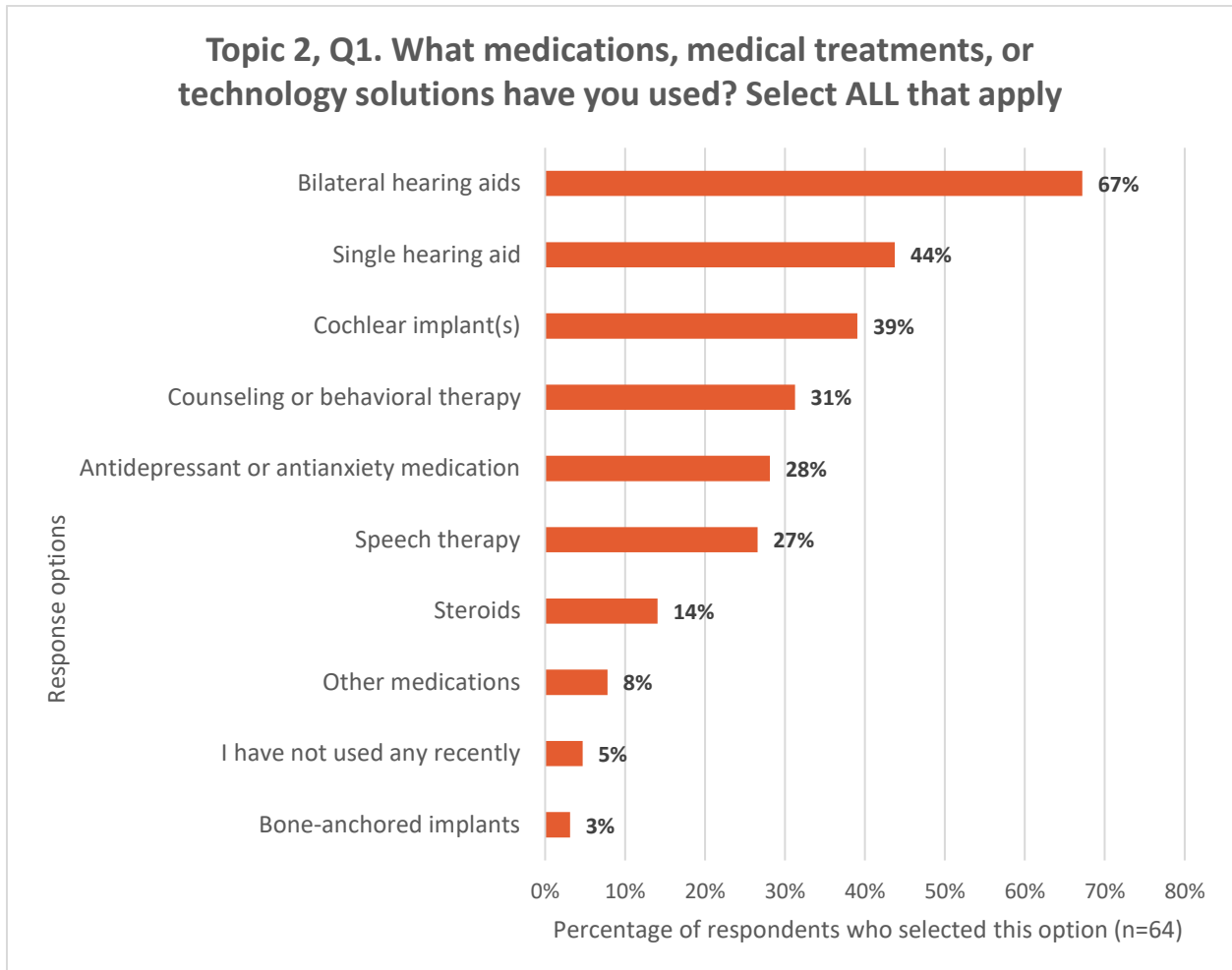


Voice of the Patient Report: *Living with Sensorineural Hearing Loss*

Appendix 6: Topic 2 Polling Results. Current and Future Approaches to Treatment for Sensorineural Hearing Loss

These graphs include those attendees who chose to participate in online voting. The number of patients who responded to each polling question is shown below the X axis (n=x).

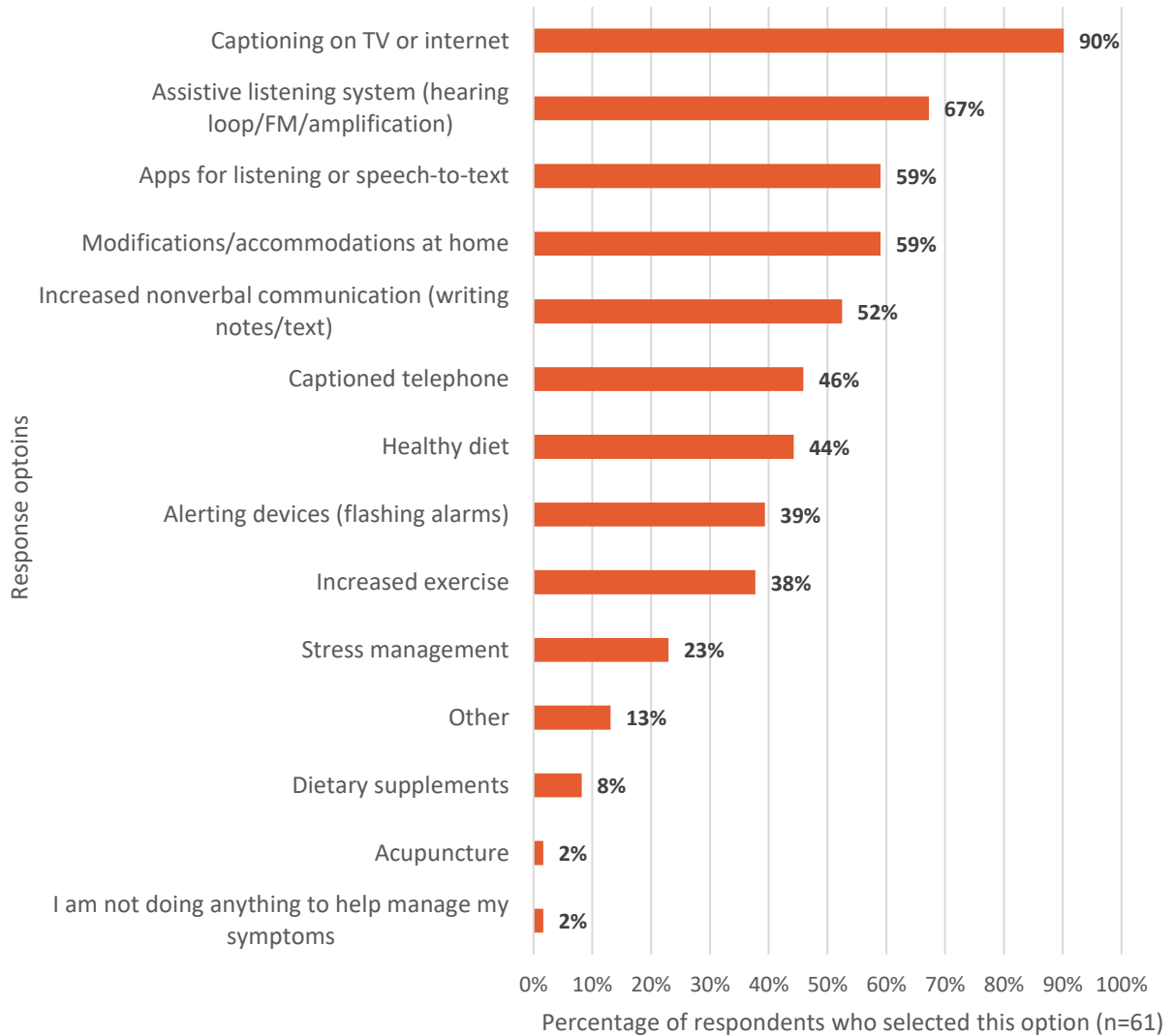
The response rate data for these polling questions are not considered scientific data but provide a snapshot of who participated in the EL-PFDD meeting and are meant to complement the live and pre-recorded comments throughout the meeting.



Voice of the Patient Report: *Living with Sensorineural Hearing Loss*

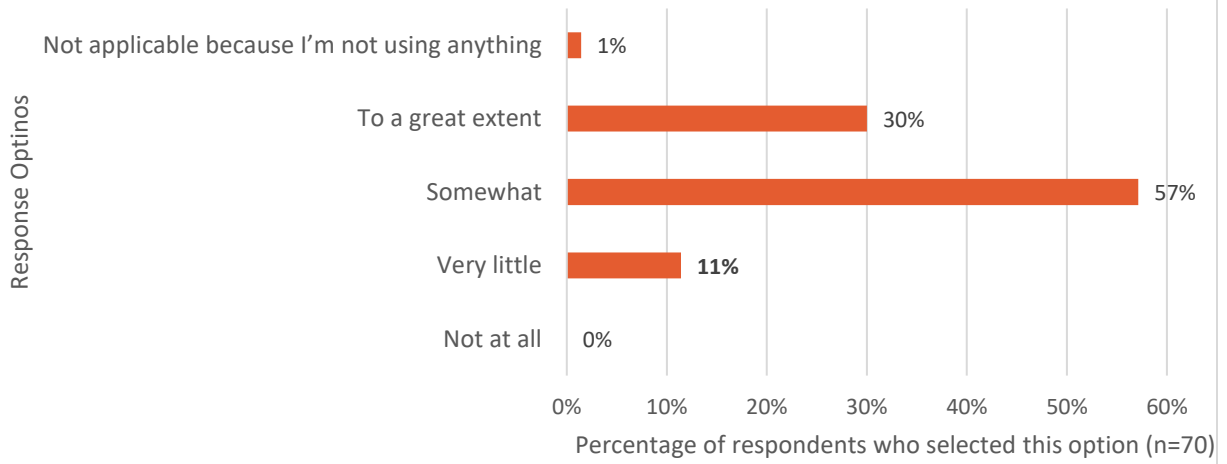
Topic 2, Q2. Besides medications and treatments, what are you currently doing to help manage the symptoms of hearing loss?

Select ALL that apply

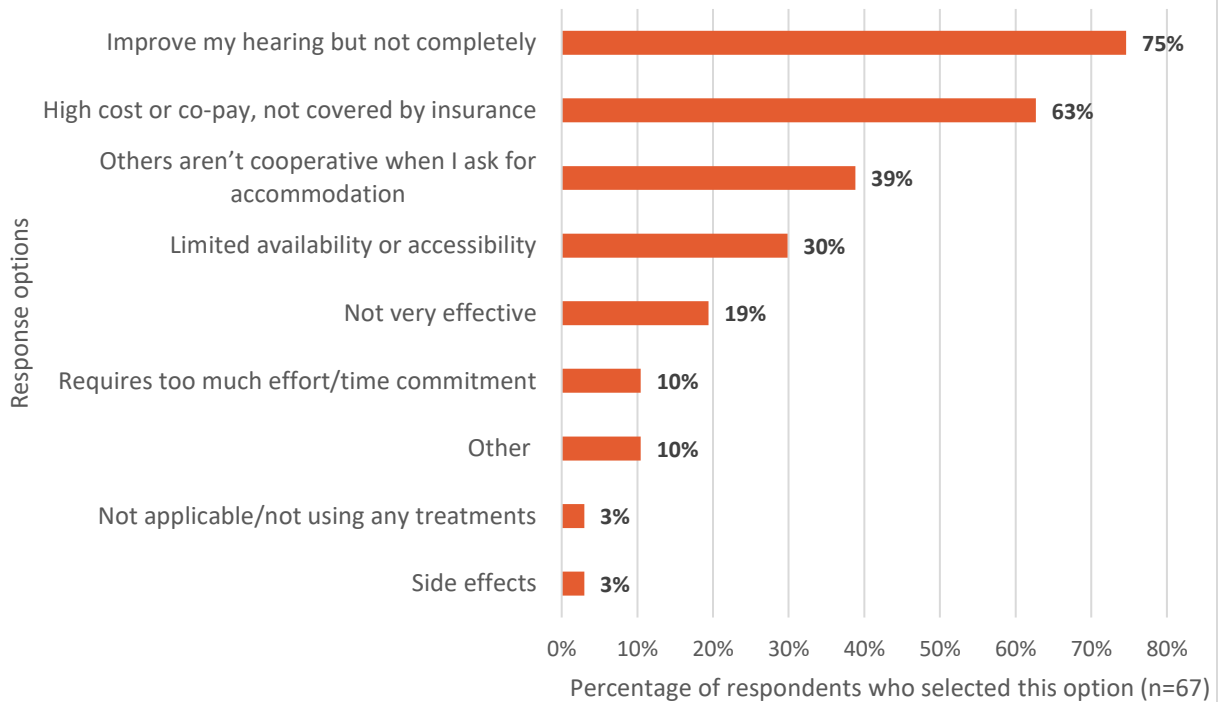


Voice of the Patient Report: *Living with Sensorineural Hearing Loss*

Topic 2, Q3. How well does your current regimen control your condition overall?



Topic 2, Q4. What are the biggest drawbacks of your current approaches? Select up to three



Voice of the Patient Report: *Living with Sensorineural Hearing Loss*

